Clinical Auscultation of the Heart

By

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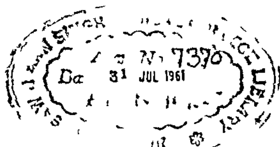
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TO

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ROSALIND AND IRMA



Preface to the Second Edition

Ten years have elapsed since the first edition of this book was published. During this period interest in auscultation has increased tremendously. There was a time, several decades ago, when the stethoscope had fallen into partial disrepute. Without deprecating the great value of symptoms and history taking in appraising the cardiac status—especially as they pertain to the early diagnosis of congestive heart failure or angina pectoris—it has become quite clear that a few minutes of intelligent auscultation of the heart can reveal many findings of vital importance.

The past decade has afforded the authors an opportunity to increase the scope of this book. Numerous additional auscultatory phenomena have been observed, studied and illustrated, examples of which were not available or not known previously. Greater experience with congenital heart disease has broadened the knowledge of the significance of certain murmurs and sounds, especially splitting of the second sound. Variations of the physical findings in different patients suffering from the same underlying condition have often necessitated the inclusion of more than one illustration of the auscultatory phenomena in question.

In general, it has become clear that, despite the aid and the advances that have come from the newly introduced more specialized techniques of cardiac investigation, the value of the simple stethoscope is steadily increasing.

The Authors

Without this original suspicion derived by the stethoscope, these other tests would not be performed and the correct diagnosis will be entirely overlooked In some instances, errors that could have been avoided by intelligent auscultation have been matters of life and death For the above reasons it appeared timely to discuss in detail the simple data pertaining to bedside auscultation that can be grasped and applied by any physician in the practice of medicine

In addition to the ausculatory findings, it seemed that the discussion would become more vivid and useful if some comments were made concerning the related clinical conditions and the therapeutic implications involved These points have been taken up and have been illustrated by occasional specific experiences and case reports

We wish to express our indebtedness to the house staff of the Peter Bent Brigham Hospital for the assistance given in calling our attention to the various cases displaying peculiar acoustic phenomena Without their help many of the uncommon illustrations displayed here would not have been obtained We are also grateful to the undergraduate students whose perplexing questions often served as a stimulus to present graphic proof of disputed points This has already been of great value in the teaching of auscultation of the heart. Finally, we want to thank Dr Harold D Levine for performing the arduous task of constructing a suitable index

THE AUTHORS

Acknowledgments

MANY individuals have helped us in the preparation of this Second Edition. To each we would like to express our sincere appreciation, but first, to the students at Georgetown University School of Medicine, to the House Staff and to the Fellows in Medicine, Division of Cardiology, Georgetown University Hospital. Their continued interest in auscultation of the heart, their calling our attention to specific examples on the wards, and their pertinent questions have been a real stimulus in revising and enlarging the scope of this book.

Special thanks are due Doctors Recep Ari, Michael A. Corrado, and Jack P. Segal, *presently on the teaching staff of* Georgetown University Hospital, who have continued to demonstrate their unusual interest in this aspect of cardiology. To Dr. Joseph K. Perloff, in particular, we are most grateful. His help and his continuing investigations of the physiology of heart sounds and murmurs has added greatly to our understanding. Dr. Charles A. Hufnagel, Professor of Surgery, has always allowed us unlimited opportunity to study his patients, and has provided us with the surgical correlation in many. For this cooperation we are most grateful.

Some of the patients have been at sister institutions, and we have kindly been allowed to study and record them. We wish to thank the following: Doctors Robert Grant, Andrew G. Morrow, Eugene Braunwald and Samuel Fox, National Heart Institute, Bethesda, Maryland; Brigadier General Thomas Mattingly, MC, USA (Ret.), former Chief of Cardiology, Loren Parmley, Lt. Col., MC, USA, and Robert Hall, Lt Col., MC, USA, Walter Reed Army Medical Center, Washington, D.C ; R J. Pearson, Captain, MC, USN, Chief of Cardiology, Gerald I. Shugoll, Lt , MC, USN, William Jacoby, Lt. Comdr., MC, USN, Bethesda Naval Hospital; John Evans and Irene Hsu, George Washington University Hospital, Washington, D.C.; John Boone and Dale Groom, University of South Carolina; Robert Gilston, Amsterdam, New York; Ike Muslow, New Orleans, La.; Frank Marcus, Georgetown University Hospital, Washington, D.C.; Dr. John Staple-

Chapter 3

Introduction

MANY STUDIES have been made of the heart sounds in normal and pathologic conditions using various types of techniques. In recent years most of these have consisted of photographing the sounds electrically and correlating the events with simultaneous electrocardiogram and mechanical tracings obtained from the apex beat and jugular pulse.

Harvey, as early as 1628, mentioned that heart sounds were audible, though no doubt others must have heard the heart beat by direct application of the ear to the chest many years before. It was not until the time of Laennec (1819) that we had a precise description of the heart sounds in normal and abnormal states. Later, the subject was more extensively studied by Potain (1866). The introduction of the capillary electrode by Einthoven (1894) and the segment capsule by Frank (1904) further extended our knowledge of heart sounds and other vibrations. Later still the string galvanometer, and finally the amplifier tube apparatus, were used to photograph heart sounds.

Throughout these years the origin of the normal and abnormal heart sounds has been a matter of great contention and theoretical speculation. Even at present there are differences of opinion concerning many of the disputed points It is not the purpose of this discussion to analyze the exact mechanism of the acoustic phenomena. Publications are available that go into the details of phonocardiography, notably those of Cossio, Orias and Braun-Menendez. In this country, the studies of Wiggers, Wolferth and Dock have added a great deal to our knowledge of the significance of heart sounds. However, there is need for a discussion of the various types of heart sounds and murmurs that one can readily hear, to illustrate what deductions may be made from changes in the character of the sounds, how the various cardiac arrhythmias may be recognized, what inter-

pretation can be made from different murmurs, and what significance other peculiar sounds may have

Sounds that can only be detected by a sensitive phonocardiograph and not by the human ear using the ordinary stethoscope, may have certain meaning, but will not be taken up here In point of fact, in the present state of our knowledge, it is unlikely that sounds which the average ear cannot hear would have any clinical significance except in very rare instances Our attention, therefore, will be confined to the exposition and clinical interpretation of the sounds that the ear can hear Phonocardiograms are used in this book to portray *visually only what we have heard clinically* with the stethoscope

The method to be employed will consist of discussing various acoustic phenomena, giving the clinical significance of the particular finding, and then illustrating the point involved by actual sound tracings obtained from patients In these illustrations, simultaneous electrocardiograms (generally of Lead II) and phonocardiograms will be presented It is realized that exact correlations as to time cannot be made between the events of the electrocardiograms and those of the heart sounds More accurate relationships could have been made if simultaneous tracings had been taken from the jugular pulse and apex beat This would have been much more difficult and it would not have aided in the clarity of the clinical discussion, nor would it have added to the practical use to which these illustrations may be put However in some of the phonocardiograms the carotid arterial pulse tracing is utilized particularly when portrayal of aortic valve closure is desirable (e g, splitting of heart sounds)

In reproducing heart sounds by the phonocardiographs in current use, one can increase or decrease the volume of sound and hence the amplitude of vibrations that are to be photographed A difficulty arises in that, with greater amplification extraneous sounds may appear When the actual heart sound to be investigated is quite faint, the sensitivity of the apparatus must be increased to those very high intensities at which extraneous sounds may appear The graph that will serve the best purpose for elucidation of what is audible by the average physician is one that will show no vibration whatever during the intervals when the human ear can hear nothing and yet will clearly indicate faint sounds that are audible However, the sensitivity of the apparatus should be such that definite vibrations coming with any degree of regularity should not be attributable to artefacts but to the action of the heart or other recognizable mechanisms A certain amount of experience is required to obtain suitable graphs and to be able to recognize outside or confusing factors, such as the effects of rubbing of the skin breathing coughing or noises made in the room or by the machine itself

The reader should not get the impression that recordings of heart sounds are necessary in general practice They are used here mainly

to illustrate what has already been heard and to help visualize the points taken up in the discussion. The situation is much the same as the use of the electrocardiogram in the detection of cardiac arrhythmias. The more one has studied electrocardiographic tracings in relation to bedside findings, the less he needs such tracings in the diagnosis of irregularities of the heart. Likewise, it is hoped that the analysis and study of these phonocardiograms will enable the reader to draw the proper inferences without such records.

An erroneous impression that still exists in the minds of some is that the taking of a phonocardiogram will immediately settle any point of discussion concerning a disputed auscultatory finding. For example, one observer may state that he hears the diastolic rumble of mitral stenosis. Another may be unable to hear this murmur. In an effort to arbitrate this particular dissension of views, a phonocardiogram may be taken. If the diastolic rumble is not portrayed, this does not necessarily mean that it was not present. If the microphone had not been placed at the exact spot, using light pressure, or the breath held in the correct phase of respiration, and the record taken after a maneuver to "bring out" this murmur, it would not be recorded on the phonocardiogram. All too frequently we have heard the statement that "the murmur was not there because it was not shown on the phonocardiogram." On the other hand, a phonocardiographic finding of the auscultatory phenomena in question may settle a dispute, or may be valuable in timing a sound or murmur.

At the outset it may be well to clarify a few fundamental points about auscultation. In order that a sound may pass the threshold of audibility, it must have a certain intensity, frequency and duration. If it is not loud enough, if its frequency is too slow or too rapid, or if it does [illegible] heard. Different individuals differ [illegible] Sounds of different frequencies o[illegible] laws, so that an appropriate filtering apparatus may be useful to be able to dampen some sounds, e.g., those of low frequency, in order to bring out others of high frequency, otherwise difficult to identify. Actually, the physician uses his stethoscope as a filter. The bell portion, applied with light pressure, accentuates low frequencies (such as a diastolic rumble) and filters the high frequencies; whereas the diaphragm, with firm pressure, potentiates the high frequency (such as an aortic diastolic blow) and diminishes or eliminates the low frequencies. Furthermore, the sounds produced within the chest are transmitted through varying thicknesses of tissue before reaching the observer's ears. The efficiency of sound transmission is greatest in solid, less in liquid, and least in gaseous media. We therefore can only infer what is going on within the heart from sounds heard on the surface of the chest, by considering the condition and configuration of the lungs and chest wall.

In the total evaluation of a patient with possible heart disease, it

is generally agreed that the history is the single most important aspect Despite the numerous advances that have taken place, including such specialized techniques as cardiac catheterization, angiocardiography, ballistocardiography, electrokymography, electrocardiography and roentgenology of the heart, the physical examination still ranks as the second most important of the individual aspects of cardiovascular evaluation The teaching of auscultation of the heart is still inadequate in our medical schools today Although the simple stethoscope still remains one of the most important tools available for the diagnosis of heart disease, the majority of students and physicians at present are not making proper use of it It goes without saying, however, that one should not attempt to assess the cardiac status of any individual without utilizing all of the facets of the cardiovascular evaluation, including history, physical examination, electrocardiogram, x ray and other laboratory examinations whenever necessary

Specific information, invaluable in the diagnosis and treatment of the individual patient, is obtained from stethoscopic examination Never before in the history of medicine has clinical auscultation of the heart been of more importance than it is today In fact, interest in the whole subject appears to be greater now than at any previous time Contributing to this is the great recent progress in the field of cardiovascular surgery In this field the diagnosis of conditions such as mitral stenosis, patent ductus arteriosus, arteriovenous fistula and others depends to a very great extent on the auscultatory findings As examples one could mention the diastolic rumble of mitral stenosis and the typical machinery murmur of patent ductus arteriosus Without these typical findings, one would hesitate to make the diagnosis of either of these conditions In addition, the finding of a diastolic gallop rhythm may be the first evidence of cardiac decompensation or of serious heart damage The classic friction rub would immediately establish the diagnosis of pericarditis the typical bizarre crunch crackling sound characterizing mediastinal emphysema would instantly suggest a condition that otherwise might be confused with diseases such as acute myocardial infarction or pericarditis Numerous other examples could be cited to illustrate the importance of auscultation in the diagnosis and treatment of heart disease today Unfortunately, however, even at the present time the stethoscope is not utilized to its fullest extent Inferior and inadequate stethoscopes are still frequently employed, and many simple principles involved in the proper use of the stethoscope are not yet appreciated

THE STETHOSCOPE

Type of Stethoscope Many physicians buy only one stethoscope in a lifetime Care should therefore be taken in the selection Each of

us becomes used to his own stethoscope and frequently finds that he does not hear so well using others. Rappaport and Sprague investigated acoustic principles involved in auscultation of the heart and emphasized the importance of incorporating in the stethoscope the correct tubing, earpieces, bell and diaphragm. The effect of varying the amount of pressure with which the bell and diaphragm are applied to the chest wall is also stressed. We have included these principles in the following discussion.

KEY TO ALL FIGURES

S_1—First heart sound
S_2—Second heart sound
S_3—Third heart sound
G—Gallop sound
A—Atrial sound
X—Artefact or extracardiac sound
S_4—Opening snap of mitral valve
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"—Fuzziness of base line
SM—Systolic murmur
DM—Diastolic murmur
PM—Presystolic murmur
S—Sound heard with blood pressure auscultation
P—"Pistol-shot" heard over peripheral arteries with aortic insufficiency
L.S.B.—Interspace at left sternal border
A.G.—Atrial gallop
V.G.—Ventricular gallop
E—Ejection sound
A_2—Sound of aortic valve closure
P— Sound of [illegible]
[illegible]

One's stethoscope should be capable of detecting both the low and high frequencies of heart sounds and murmurs. To do this, both bell and diaphragm chest pieces are usually necessary. It is of utmost importance to use both the bell and the diaphragm in examination of all patients. The bell piece is designed to accentuate the lower frequencies of heart sounds by filtering out the higher frequencies. It is particularly applicable for hearing the rumble of mitral stenosis or a faint diastolic gallop rhythm. On the other hand, a blowing type of systolic murmur, where the f[illegible] better w[illegible] murmur [illegible] murmur of pulmonic insufficiency are good examples of such sounds. In many instances, the aortic diastolic murmur would probably be overlooked if only the bell were used, but might readily be heard with the diaphragm.

Importance of Pressure When Applying the Stethoscope to the Chest. The amount of pressure with which the stethoscope is applied to the chest is one of the least stressed aspects of adequate auscultation. For example, if one has a patient with rheumatic heart disease and combined mitral stenosis and aortic insufficiency, he will hear the diastolic rumble best *by using the bell chest piece and holding* the stethoscope quite lightly—barely touching *the skin in a localized*

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C—Click sound

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spot over the apex In case the diastolic rumble is faint (grade I or grade II), it could easily be overlooked, even if the bell were used, unless very light pressure is applied Even with average pressure, this particular murmur will often be missed, or it might not be heard if the diaphragm alone were used On the other hand, for detection of the high pitched diastolic murmur of aortic insufficiency, the diaphragm is best utilized A grade II early, blowing diastolic murmur of aortic insufficiency would probably be missed if one used just the bell, but it is immediately detected with the diaphragm When the murmur is grade I or II, it may be overlooked even with the diaphragm unless the stethoscope is firmly pressed against the chest wall (Often one must exert enough pressure so that the skin shows the imprint of the stethoscope on the patient's chest wall By exert-

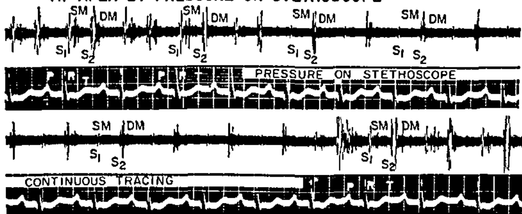


FIG 1 Boy age 18 who had a traumatic rupture of aortic valve and interventricular septum after being kicked in the chest by a horse Note decrease in sounds and murmurs particularly the first sound (S_1) and systolic murmur (SM) when pressure was applied to the stethoscope

ing significant pressure, low frequency sounds are diminished and the high frequency components are accentuated, making this type of murmur more readily heard For this reason, other high frequency sounds such as friction rubs are usually best heard by applying firm pressure with the diaphragm Firm pressure with the stethoscope often greatly diminishes or causes the disappearance of an apical systolic murmur Figure 1 shows an example of this A blowing diastolic murmur of aortic insufficiency was also present which although decreased slightly in intensity had its high frequency components emphasized

Another good example of the necessity of varying degrees of pressure is the gallop rhythm Most diastolic gallops are of low frequency and are faint No difficulty is experienced hearing a loud ventricular diastolic gallop, but faint gallops of this variety, which are the most common, are frequently overlooked In order to bring out this faint

low frequency sound, the stethoscope must be applied with very light pressure, barely touching the skin. The fainter gallops of this type are poorly heard using the diaphragm.)

Tubing. Tubing is often of the wrong diameter, too long, torn and poorly fitting. The length of tubing makes considerable difference in how well one hears the various auscultatory phenomena. Actually, the shorter the tubing and the closer the ear to the heart, the better. However, one must achieve a practical solution of the problem, and tubing varying from 10 to 15 inches in length has proved perfectly satisfactory.

The importance of the length of stethoscope tubing was well demonstrated on one occasion when we were making cardiac rounds. One patient had rheumatic heart disease with a moderate degree of aortic insufficiency. The early, blowing, aortic diastolic murmur was easily heard and was loudest with the patient sitting, leaning forward, and holding his breath in expiration. Firm pressure was exerted on the diaphragm chest piece. A visiting physician started to examine the patient, and, as he reached for his stethoscope, we were amazed at the length of the tubing. One end of the stethoscope was in the inside pocket of his coat on the right side, and, after he had removed this portion, a lengthy serpentine tube encircled him under his coat and connected with the other end in the left rear pocket of his trousers. The length of the tubing must have been approximately three feet. After he had listened to the patient, we repeated our examination using his stethoscope. The heart sounds were audible, but the typical, early, blowing diastolic murmur previously described could not be heard. We then asked our visitor why he used a stethoscope with such long tubing. He stated, "In my country I have contact with a large number of patients who have tuberculosis, and for this reason I keep a good distance from them." We all laughed with him at this remark and agreed that, for his purposes, the longer tubing was not a bad idea, although for adequate auscultation of the heart and lungs, important findings would necessarily be overlooked.

The importance of the diameter of the lumen of the tubing should likewise be emphasized. It has been determined that a diameter of approximately ⅛ inch is the most satisfactory.

The Diaphragm and Bell. Not infrequently the diaphragm of the stethoscope becomes cracked. Its further use is to be discouraged, and a replacement should be obtained as soon as possible. A too-rigid diaphragm will diminish the intensity of all sounds, and too flexible a diaphragm will accentuate the low pitched ones. Occasionally, one encounters a physician using no diaphragm whatsoever after this piece has been broken. In such an instance, adequate auscultation is impaired for the higher frequencies, such as those of aortic insufficiency and friction rubs. Another not uncommon practice is to replace a broken diaphragm with x-ray film, a rubber covering or a similar type of material that occasionally accompanies medical ad-

area Thus thrombosis of the iliac and femoral veins causes enlargement of the entire leg, whereas a failure of the super ficial venous return, as in varix, and even an extensive throm bosis of varicose veins, leads to almost no swelling

Such gross distinctions as these are clear enough, and if all circulatory disorders fell into these categories, they would very readily be understood Unfortunately there are arterial diseases which are not organic but functional, not perma nent but spasmodic—disorders of vasomotor control, whether through the sympathetic system or chemical mediation, un natural reactions to the emotions and especially to cold, such as Raynaud's disease There are, also, combinations of ar terial and venous occlusion, notably thrombo angiitis oblit erans (Buerger's disease) There are arterial spasms apparently reflex in nature and secondary to a variety of wounds and injuries Some of these are temporary, others are prolonged and associated with serious changes in skin, bones, and joints —painful states, as a rule, including the causalgias There are even arterial spasms secondary to, or at least associated with, venous thromboses Finally, there are inexplicable and perma nent states of vasomotor irritation causing cyanosis, cold ness, sweating, and, in some instances, ulceration of the extremities

The arterial aneurysms and venous malformations, such as nevi and the arteriovenous fistulas both congenital and trau matic, also affect the limbs, though since they exhibit them selves in other places they are by no means peculiar to these parts A brief account of them, if only for purposes of com parison, will therefore be included here

The Arms and the Legs —There is a distinction of some importance, rather helpful in diagnosis, between the diseases of the arms and the legs Whether because the blood pressure is higher in the lower limbs than the upper or because of the wear and tear due to a more violent use, the legs commonly show the effect of arteriosclerotic stiffening and endarteritis, whereas the arms almost never do so This is true both of the chronic narrowings and acute thromboses The combined

thrombosis of both arteries and veins in thrombo-angiitis obliterans is also far more troublesome to the legs than the arms, though here the distinction is much less clean-cut than it is in the case of the purely arteriosclerotic disorders. The legs and pelvis are also the seat of most of the venous thromboses and of the unusual secondary arterial spasms related to them.

The arms, by contrast, tend to be the usual site of the many states of arterial spasm brought about through the mediation of the vasomotor government in general. To all generally distributed reactions the arteries of the legs of course respond, though in a less noticeable and troublesome way than do those of the arms. But there are certain states, such as the rare arteritis, associated with irritation of the brachial plexus by a cervical or first rib, and the effects of certain injuries of the great nerves, causalgia or causalgia-like states, which are almost peculiar to the upper extremities.

There is also a distinction between states of spasm and organic disease, not to be taken too seriously but helpful in diagnosis. The area principally subject to the effects of *peripheral* arterio-spasm occupies little more than the feet and hands. That is to say, it is limited centrally and sometimes with almost a glove- or shoe-like abruptness shortly above the wrist and the ankle. Thus the diseases which show themselves in changes confined to such areas will most often have a predominantly vasospastic background. See, for instance, a youngish man, a heavy smoker, suffering from a pronounced intermittent limp and find, on passing your hand down his legs, that cool skin is encountered rather abruptly at almost the level of the shoe-tops. Whatever degree of organic obliteration he may exhibit, you may properly judge that there is also an element of sympathetically controlled arterial spasm in his case.

This rough analysis of the vascular diseases of the extremities will serve as the *outline* for this book. Information about them is increasing and at such a pace that classification is apt to be inadequate and explanation fallacious. However, there is here outlined a sort of index of disabilities which may be

useful in pigeonholing the various circulatory disorders

Arteriosclerotic Deficiencies
  Diabetic Infections and Gangrene
Thrombo-angiitis Obliterans
Vasospastic Disorders
  Raynaud's Phenomenon
  Vasospasms, acute traumatic
  Vasospasms, chronic and related to injury and infection
  Vasospasms, functional, permanent and unclassifiable
Arterial Embolism
Varicose Veins and Ulcer
Thrombophlebitis and Venous Embolism
  Post phlebitic States
Arterial Aneurysm and Abnormal Arteriovenous Communications
Lymphedema, Lymphangioma and Elephantiasis

The most significant function of the circulation is the nourishment of the tissues At first sight it might seem that this function, in the case of the limbs, is quite the same as that performed for the rest of the body Such is hardly the case Though it is true that when at rest the limbs make little demand upon the circulation, in action their demands are enormous Everyone is familiar with the slowing up of the athlete, the giving out of his legs, in the late thirties His heart is probably as good as ever His endurance, partly owing to his increased experience and skill, is probably greater But his elasticity, as opposed to his capacity for sustained effort, that is, his capacity for muscular response at high speed, is beginning to lessen The cause of this change lies presumably in the failure of his arterial system to respond actively enough to the call made upon it by the muscles Carry this functional deficiency a little farther and you have a basis for the intermittent limp At rest, the legs of the individual whose arterial system is becoming limited are comfortable In action, they are insufficiently supplied with blood, and the characteristic intermittent painful numbness sets in Carry the circulatory

deficiency still farther, combine it with minor injuries and the cooling of small parts with large surfaces, and the extremities become more than functionally ill-nourished. They are actually liable to necrosis—senile or presenile gangrene.

The second function, and one upon which familiar observations and tests depend, is the maintenance of the surface temperature. This of course is only a part of the control of body temperature in general. However, the local cutaneous temperature is of chief interest here. Warmth of the skin is a sign of an abundant blood supply. Coldness is an evidence of a deficient circulation. This is simple enough. But a deficient circulation may be restricted either organically or merely by functional contraction. And so a distinction must be made between these two states, a matter of deciding whether or not the arterial system, especially the finer parts of it, are capable of contraction and expansion. An organically deficient arterial supply makes a relatively cold surface but above all is so inelastic as no longer to respond to a reflex call for temperature regulation. Thus the skin of a limb tends to take on, not the deep temperature of the body but the temperature of its environment. It can slowly be warmed by the surrounding air, and rather more readily be cooled. A normal set of vessels responds to a hot environment, and particularly to any signal received from the heat-regulating centers, by dilatation with consequent warming and flushing of the skin in an effort to disseminate heat, the medullary centers being extremely sensitive to *any* rise in the temperature of the blood. By the same token, normal vessels respond to external cooling by contraction, lest heat be lost through radiation. Now just as stiff, contracted arteries fail to respond to such influences, unnaturally irritable vessels may respond too readily. A little cooling of the environment, a little nervousness or fear, sends them into a state of spasm, whereby the skin becomes pale (or blue) with cold. This is of course a gross picture. The normally and overly responsive small arteries are spoken of as if their reactions were only to nervous control of body temperature in general. As a matter of fact they respond to local stimuli

as well By warming a foot, for instance, its small vessels can be made to dilate quite apart from any general influence

The large and small vessels do not react, respectively, in quite the same way to contracting and dilating influences Vasodilatation and vasoconstriction chiefly occur in the small arteries and arterioles Thus it is upon the surface, particularly in the feet and hands and above all in the fingers and toes, that changes in surface temperature are most sensitively displayed Sir Thomas Lewis has revealed the purpose of the numerous fine arteriovenous anastomoses, muscular and richly innervated, which are present upon the surface of the body and especially in the digits, the palms and the soles *These, when dilated, permit a flood of arterial blood to warm* the skin Similarly, their contraction aids in cooling the surface Thus, as compared with the body and the more bulky part of the limbs, the feet and hands are far more rapidly warmed and cooled and quickly reveal their state in the temperature of the skin. It is, in fact, in the fingers and toes that *peripheral vasoconstriction and vasodilatation can most easily* be studied

The Sympathetic System, previously mentioned but not actually described, now requires consideration It is an outflowing mechanism, exercising vascular control and, by the stimulus of pain, rage and fear, energizing the body for action Shakespeare might have been collaborating with Walter Cannon when he wrote

> "In peace there's nothing so becomes a man
> As modest stillness and humility
> But when the blast of war blows in our ears,
> Then imitate the action of the tiger
> Stiffen the sinews, summon up the blood,
> Disguise fair nature with hard favored rage
> Then lend the eye a terrible aspect,
> Let it pry through the portage of the head,
> Like the brass cannon, ***
> Now set the teeth, and stretch the nostril wide,
> Hold hard the breath, and bend up every spirit
> To his full height!" *Henry V* Act III, Scene 1

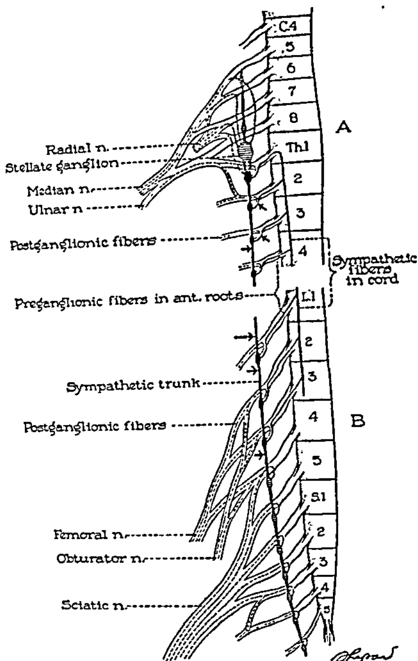


FIGURE 1. A DIAGRAMMATIC SKETCH OF THE SYMPATHETIC INNERVATION OF THE LIMBS. The sympathetic ganglia and chain, solid black; the preganglionic rami, solid black; the postganglionic rami, dotted lines. Arrows indicate points at which the rami and chain should be interrupted to secure preganglionic denervation Interruption above L, 1 and removal of L, 1, 2 and 3 gives vasodilatation as high as the thigh but is only to be practiced in females.

In other words, the sympathetic system, stimulated by any excitement, raises the blood pressure, constricting the arteries, and drives the blood to the muscles which it abundantly furnishes with immediately usable fuel But in the meanwhile, it stops all processes not immediately required for action By causing the peripheral vessels to contract, it halts all loss of heat, at the same time turning the surface pale, dilating the pupil and widening the eye, and by an action usually but not always consistent with this vasomotor response, it erects the hairs and sets the skin to sweating By contrast, paralysis of the sympathetic leaves the muscular arteries relaxed, the skin flushed and dry, the pupils contracted In the normal, balanced state, the sympathetic, of course, maintains a steady vasomotor tone

All this is accomplished by a system consisting of a series of nerve cells and relay stations From cells in the very oldest part of the brain, fibers descend the cord and pass out with the anterior spinal roots to make contact with secondary cells in the long, paravertebral ganglionated chain The little nerves which carry these fibers from the cord to the ganglions are the preganglionic or white rami Through every one of these, several ganglia are activated Next, from the cells in the ganglia, other fibers pass on to be distributed via the peripheral nerves to the blood vessels and sweat glands all over the body These are the gray or postganglionic rami There are other way stations to the various thoracic and abdominal viscera, but the simple system just described serves for the blood vessels In Figure 2 the distribution of sympathetic fibers to the arterial system of the limbs is pictured It will be noticed that the fibers flow out upon the great vessels from the principal nerves at a series of levels but do not travel far along them, and that the distribution of the sympathetic supply from the principal nerves of each limb corresponds to their respective sensory fields upon the surface

To this description it is only necessary to add that the sympathetic supply to the arm arises in the upper thoracic cord, passing out by white rami to the second and third thoracic

ganglia and along the sympathetic chain to the stellate ganglion (first thoracic and inferior cervical); thence, principally from cells in the stellate, in gray rami, to the brachial plexus. The supply to the leg comes from the lower thoracic and lum-

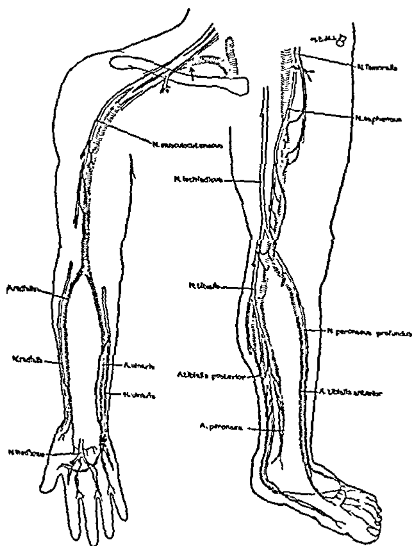


FIGURE 2. THE DISTR... BLOOD ... and Po ... point at which fibers destined for the brachial artery may be exposed to irritation as they pass over the highest rib.

bar cord Some preganglionic fibers pass in white rami to the second and third lumbar ganglia, and from these ganglia a number of postganglionic fibers go out to the vessels of the thigh But most of the preganglionic fibers descend in the sympathetic chain to ganglia below the second and third lumbar, where they make their contact with cells whose postganglionic fibers are distributed, via the sciatic to the principal arterial system of the legs This arrangement permits division of the preganglionic rami to both upper and lower limbs without injury to the cells of the postganglionic fibers Thus, stimuli from the central nervous system can be cut off* from the vessels of the limbs without loss of the *local* postganglionic governing mechanism This feature will be developed in the story of the operative treatment of vasospasm (Chapter IV) and is a fundamental consideration in present-day vascular surgery

In the foregoing paragraphs the vasomotor nerves have been described as if they consisted only of vasoconstricting fibers About outgoing vasodilating fibers, it such exist,† less seems to be known Vasodilatation takes place not only when central vasoconstrictor control is artificially lifted but under the influence of the products of local metabolism which cause the blood supply to fluctuate very delicately and accurately according to the needs of the tissues The blood supply of the muscles is governed in this way If the circulation to a given area of muscle is arrested, sufficient vasodilatation occurs in

* It is confusing to discover as Oughterson, Harvey and Richter have done, that after an apparently complete sympathetic denervation of the lower limb some vasomotor control yet remains (posterior tibial nerve block causes additional vasodilatation) Their experimental observations suggest that vasoconstrictor fibers reach the sciatic nerve from sources in the spinal cord lower than the lowest hitherto recognized exits for sympathetic nerves (that is caudad to the third and even the fourth lumbar roots)

† In the normal individual they have never been proved to exist However, Lewis and Pickering have shown that in anyone suffering from vasospasm as in Raynaud's disease *full* vasodilatation in the little finger as called forth by heat is prevented by blocking with procaine the corresponding ulnar nerve Yet blocking the ulnar nerve abolishes all vasoconstriction in the ulnar field paralyzing vasoconstriction fibers Thus it would seem also to paralyze vasodilator fibers A parallel phenomenon has been found true in the leg

that area so that when blood flow is again released its volume is greatly increased until the oxygen debt is paid.

The statement has also been made that sympathetic nerves do not travel for any great distance upon large arteries. This does not deny the fact that stimuli of some sort—possibly in-going sensory impulses, for arteries appear to be sensitive —do travel along the great vessels. The fixation of an embolus in the femoral artery, for instance, or an inflammatory reaction in its wall may be associated with peripheral spasm in the area served by the artery. Cut out a portion of this great vessel, which Leriche likens to a long, inflamed nerve, and a vicious reflex cause of vasospasm is at once relieved, a reaction which usually improves the circulation in the peripheral field. Whether that division has succeeded because of interrupting in-going or out-going impulses is still a question. *Apparently* thrombosed or inflamed veins occasionally excite a similar reflex vasospasm. Injured nerves are capable, under unusual circumstances, of setting up a somewhat similar symptom complex. That such states involve in some way the sympathetic system is beginning to be made clear by the relief secured from a temporary or permanent block cutting off all the sympathetic impulses to the part affected. However, as will appear when these states are discussed, the mechanism in most instances is not only difficult to understand but is not necessarily of the same sort in different cases. All that can be affirmed is that a persistent vasospasm, even of a large arterial tree, can take place *as a result* of a considerable variety of stimuli affecting not only the arteries themselves but the great veins and nerves as well.

Vascular Exercise and Reactive Hyperemia.—In the previous paragraphs some account has been given of the normal and pathological physiology of vasoconstriction in so far as it affects the limbs, and the suggestion has been made that vasodilatation, the reverse of that picture, occurs, not as an independent process, but as one which takes place when vasoconstriction is abolished, as by fever or as a reaction after temporary arrest of the circulation. That is, vasodilatation

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in his historical account of the procedure. And indeed Bier seems to have used venous hyperemia primarily to cure acute and chronic infections and not to influence the circulation alone. However, he established the facts: (1) that venous hyperemia induces a reactive hyperemia independently of nervous impulses; (2) that prolonged moderate venous stasis (many will remember their efforts to carry out his treatment!) is of greater benefit than the reaction to equally prolonged arterial constriction; (3) that reactive hyperemia is produced by severing large vessels (Leriche makes a point of this); and (4) that an accumulation of metabolites in the tissues is responsible for the local dilatation of the finer vessels (a contention of T. Lewis and others).

The most ambitious attempts to secure peripheral vasodilatation by making use of venous hyperemia have taken the form of apparatus designed to secure suction upon a limb followed by positive pressure, the two alternating in a rhythmic manner. The limb, being placed in an air-tight chamber, is subjected first to negative pressure, during which phase blood is drawn from the capillary bed into the venules and larger veins, after which the limb is emptied of blood by a brief exposure to positive pressure. Hermann and Reid maintain that the flow of blood through the smaller arteries and arterioles is thus increased and that the surface temperatures are correspondingly raised. By a very similar method, and depending upon Poiseuille's law—the volume per minute of fluid passing through a rigid tube is increased in proportion to the fall in pressure along the tube—Landis and Gibbon undertake to amplify the flow of blood through the peripheral vessels. In both cases, suction and pressure are mechanically controlled, a series of cycles being used, but Hermann and Reid use lower pressure for shorter periods, their suction being limited to 80 mm. of mercury (about diastolic level) for fifteen seconds and their pressure to 20 mm. for two seconds. By Landis and Gibbon's more intense and longer periods there may well be introduced an element of reaction to anoxemia lacking in Hermann and Reid's system. However, both

gives the impression of being a passive rather than an active process Present day literature is so full of explanations of the beneficial effect of this or that method of securing vaso dilatation as to bewilder the mind, a state of things partly due to the fact that in a growing field each investigator tends to advocate the method which he has developed and with which he is most familiar

Although for many decades attempts have been made to treat disorders of the limbs by inducing active or passive hyperemia, Cushing's application and release of a tight rubber tube (1902) seems to have been the first successful attempt to secure a reactive vasodilatation in the face of vascular spasm The idea undoubtedly was derived from the bright and rapid flush which accompanies the release of a tourniquet The tightened rubber tubing was intended to paralyze the vasomotor nerves by several minutes of complete arterial occlusion It has since been shown that occlusion of the arterial supply to a limb, even for as short a time as a few seconds, is followed, on release, by a vasodilatation which corresponds to the oxygen want created by the stoppage That the large as well as the small vessels react in this way is proved by measuring the total inflow of blood into such a limb, and the event can be counted upon, provided the arteries are elastic enough to respond Its exact cause is not entirely clear but may properly be regarded as a reaction closely related to that called forth by active muscular exercise and by local injury Whether the stoppage which is succeeded by such a reactive hyperemia is capable of harm, especially when applied to a whole limb, is another matter, but it must be supposed that unless it permanently widens the area of the vascular bed, the increased flow it brings merely balances the momentary damage done by the occlusion

Venous pressure, applied for a prolonged or short interval, is capable of inducing, less violently, a similar reaction Although the application of this principle is associated with the name of Bier, apparatus for securing venous hyperemia by suction is perhaps a hundred years older, as Hermann relates

raised, is perhaps able to carry fluids from the capillaries to tissues into which they would not otherwise be able to penetrate. In normal persons, at any rate, heightened venous pressure soon increases the flow of lymph.

The final effect of increased venous pressure, at a therapeutic (subdiastolic) level, is to lower oxygen tension in the tissues and thereby create an oxygen want, a want which must be satisfied, upon releasing the cuff, by a reactive hyperemia. It will be realized, then, that venous pressure, in addition to doing what arterial stoppage does, namely, creating in the tissues an oxygen want, raises the pressure in the capillaries while the venous hyperemia is going on and so confers whatever benefits may be received from that act.

There are yet other means of securing reactive hyperemia. The more complicated and forceful methods of doing so by vascular exercise have hitherto been described. The very simplest one is of course the Buerger-Allen system of vascular exercises, which consist simply in elevating, lowering, then actually exercising, and finally leveling and warming the limb. An *active* hyperemia can also be secured by the fever due to the injection of foreign protein, and there are moreover certain vasodilating drugs. All such have or, at least, have had their place in treatment. This will be discussed in a later section.

Testing the Arterial Supply to a Limb

Should suspicion arise that the blood supply to a limb is deficient, it is first necessary to discover whether a deficiency actually exists and, next, whether the deficiency is due to an organic narrowing of the arteries or to some temporary sort of contraction, that is, vascular spasm. Naturally, investigations in both directions will be expanded by the ambitious. Is the organic deficiency confined to one limb? Is there arterial narrowing throughout the limb, or is some particular vessel obstructed? Is vascular spasm confined to the large vessels, or is it peripheral, or general? And if the circulation is so deficient that a part of the limb must be sacrificed, how shall a

seem to induce in the smaller arteries and arterioles an increased flow of blood

Reactive hyperemia to pure venous compression is vouched for by the careful physiological observations of T Lewis and Grant (1925) More recently the clinical studies of Collens and Wilensky (who have invented an apparatus for producing intermittent venous compression) and of De Takats, Hick and Coulter (who have measured the reactive hyperemia with the aid of an oscillometer) have placed intermittent venous hyperemia upon the surgical map As a result, it may confidently be held that raising the venous pressure, as by a broad blood pressure cuff applied to the thigh, to a height just below that *of diastolic arterial pressure, say 40–80 mm of mercury,* will cause, on release, a reactive vasodilatation whose intensity is nearly proportional to the duration of the venous pressure, though actually a duration of not more than two minutes produces the best results It appears, moreover, that an interval of several minutes should elapse before compression is again applied and that a series of cycles should not last over thirty minutes (possibility of refractory phase) The reaction to venous hyperemia takes place best in a warm atmosphere

For the benefit of those who are interested in the physiological explanation of this phenomenon, it appears that during the application of the pressure, the volume of the extremity increases for some twenty seconds In this first phase, the vessels are distended and capillary pressure rises to its limit, that is, to 50–60 mm of mercury (according to Landis, the average capillary pressure is 32 mm of mercury in the arterial capillary limb and 12 mm of mercury in the venous limb) At higher venous pressures, that is, above the diastolic level, petechial hemorrhages occur and doubtless red cells escape from the capillaries into the tissues, appearing in the increased lymph stream The second phase of venous hyperemia is due to stretching of the vascular bed and tissue edema, for the filtration pressure is then increased over the osmotic pressure in the blood Thus in arteriosclerotics, whose hydrostatic pressure is often low, the filtration pressure, thus artificially

12° F. and soon the normal hand or foot responds with a rise to its natural limit, to 93°–94° F. (34.5° C.), a range of 23°–25° F.

In examining the limb, the observer will use the back of his fingers, having already applied them to his own neck to test their warmth, and will compare the patient's cutaneous temperature with his own. If in a well-warmed room the patient's skin feels warm to him, the circulation in the tested limb may be either normal or deficient. But if in a cool room the patient's foot or hand feels cold, patient and observer having been equally exposed, the patient's circulation is almost certainly deficient. Thus the simplest test is to expose the hitherto warmly covered extremities for ten to fifteen minutes to a room temperature of 70° F. or lower. If, then, the hands or feet, as the case may be, feel cold to the examiner's touch, the circulation in the patient's limbs is presumed to be insufficient. Even more striking will appear any difference in surface temperature between two symmetrically placed limbs. To take an example: An elderly individual complains of feeling a numb sort of cramp in the left calf on walking two blocks. His feet become cold in winter. He must warm them at night before he can fall asleep. He is often wakened by cramps. If he uncovers his feet and legs in a cool room—after they have become thoroughly warmed in bed—leaving them exposed for some ten minutes, both toes and feet feel *cool* to the touch. At the end of fifteen minutes the left foot is distinctly colder than the right. As the hand is passed down the leg, from knee to toes, the coolness is felt to increase from above downwards, but there is no abrupt change as the foot is approached (absence of vasomotor spasm or thrombosis of a large artery) and the foot is dry (absence of associated sudomotor excitation as a sign of sympathetic vasoconstriction). A case such as this will usually present a feeble or absent pulsation in either the dorsalis pedis or posterior tibial artery (or both) of the affected limb or limbs. The color tests usually associated with such a state will presently be described. However, there is little doubt that this is an arteriosclerotic deficiency of

safe level for amputation be determined? To carry the story of investigation further would lead to a differential diagnosis between the various known states of vascular disease It is only intended here to describe the various known means of studying the vascular capacity of the limbs Some of these means are simple, to be used on ambulatory patients and at the bedside Others are complicated and call for elaborate and often expensive apparatus For anyone willing and able to use his eyes, his fingers and his commonsense, the simpler methods afford most of the diagnostic tests he needs The more complicated methods correct and amplify the simpler ones, explain obscure signs, and furnish the accurate data needed to reveal the exact results of treatment

Ambulatory and Bedside Tests

The Temperature of the Skin—This indicates very responsively the rate of the blood flow through the limb That is, the more rapid the flow, the greater the heat lost from the surface and the warmer to the touch the skin actually feels There is a certain normal background Exposed to a surrounding temperature of about 68° F (20° C) or below, normal hands and feet tend to be cool, to feel cool to the observer's touch, having an actual temperature, as determined by special apparatus, of somewhere about 70° F, rarely much higher and never more than a few degrees lower In warm surroundings, say at 78°–80° F (about 26° C) or higher, the skin of the hands and feet tends to show a temperature of 90°–94° F (about 32°–33°C) Actually the tips of the fingers and toes offer the most striking changes of this sort, because of the very sensitive arrangement for vasoconstriction and vasodilatation which the digits possess However, the thin skin of the dorsum of the hands and feet does very well as a test surface In the influence exerted by the room temperature upon the surface temperature there is a sort of critical level or rather hill Drop the room temperature below 70° F and local circulation is slowed, the skin temperature falling nearly to that of the air about it, but raise the room temperature only perhaps 10°–

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Warm, Deep Red Skin.—Such a skin is warm because it is inflamed or because vasodilatation has been brought about by some artificial means, such as reactive hyperemia or drugs (nitrites).

Warm, Deeply Cyanosed Skin.—Unless the hemoglobin is altered (as by such a drug as sulfanilamide) such a skin is cyanosed because the circulation is delayed and warm because of external heating.

Cold, Pale Skin.—Such a skin is cold and pale because the blood flow is restricted. A slow or absent circulation gives a cyanotic tint.

Cold, Deeply Cyanosed Skin.—Such a skin is cold because the circulation is deficient or absent, and cyanosed because the circulation has been slow for so long a time that the blood has given up its oxygen. This is not inconsistent with a low-grade inflammation.

Cold, Deep Red Skin.—If the skin is very cold, the blood will not give up its oxygen and since the small surface vessels are injured and therefore dilated, the skin is deep red. Blood flow, however, is very slow.

Momentary Pressure upon the Skin.—This test, often used, is only moderately informative and may indeed be deceptive. If, for instance, the great toe is deep red, the expression of blood from the skin by pressure will be followed by a very rapid return of color. This does not indicate, if the toe is cold, a vigorous circulation, but merely the pressure at which blood is standing in the near-by vessels. In this case, a cold, red great toe would be the seat of a very slow circulation. Indeed the circulation might have ceased, yet the blush would return after pressure, as in any dependent part.

Again if a cold, already pale area is made paler by pressure, the return of color must be slow because the finer vessels in a cold skin are contracted. But this does not mean that the circulation is deficient.

The test is most valuable when applied to an elevated extremity. In the presence of a vigorous circulation, blanching due to pressure upon an elevated part is promptly followed

moderate severity but affecting the left leg more than the right

A variation upon this test of temperature can be made if the room is warm (80° F or over) The legs from the knee down are immersed in cool water, that is, considerably below the room temperature They are then dried and left exposed as before In that case, the feet and toes inadequately supplied with blood will be slow to become warm, or one leg will lag behind the other Normally, the maximum rise should be secured in fifteen minutes If, on the other hand, the room is cold, that is, well below 70° F, it may be best to start by immersing the feet in hot water at body temperature Thoroughly warmed in this way, dried and exposed, their rate of cooling can easily be discovered

Test by Color—Since color is given to the surface by the minute vessels of the skin itself, it is a less reliable test of the circulation in the limb as a whole than is the temperature However, a white skin, that is, an excessively pale one, indicates a restricted circulation A bluish skin indicates a slow circulation, one which may or may not be abundant Redness shows that plenty of blood is present, owing to inflammation or heated surroundings, which has not yet lost its oxygen The tint, in fact, as Lewis points out, must be interpreted in the light of the skin temperature For example, the cool, arteriosclerotic foot described in the previous section is pale Coldness and pallor indicate a diminished circulation, probably *not* particularly slowed But if this same foot were cold, pale, and bluish, the circulation must necessarily be not only diminished but much delayed (for since cold blood is slow to give up its oxygen it must have been long delayed to become blue and not red) As an aid to an understanding of color and surface temperature, Sir Thomas Lewis's interpretations, somewhat amplified, are here reproduced.

Warm, Pale (Pink) Skin—Such a skin is warm because for some time blood has flowed rapidly through it and pale pink because the skin is well nourished, causing vasomotor tonus moderate vasoconstriction) to be normally present

toes, the tips of which may not show color for half a minute or longer. Such a test does not relax vasospasm, for merely raising and lowering the leg fails to call forth the same degree of reactive hyperemia which sets in after the circulation to the whole limb has actually been shut off. Thus it does not differentiate organic from spasmodic obstruction. Other observations, however, may already have settled this point. If not, a somewhat more elaborate test, described by Lewis, may be used.

Heating, Elevation and Depression to Secure Reactive Hyperemia.—Though this test requires no complicated apparatus, its various steps call for strict attention to detail. The room in which the test is made should be warm. Relaxation of the blood vessels of the limb to be tested must first be secured by warming it for perhaps ten to fifteen minutes. The foot bath, maintained at blood heat, is best for this purpose. The limb is then dried and raised somewhat above the level of the body until the skin becomes pale. Thus the smaller vessels are relaxed and empty. Using a blood pressure cuff, the arterial supply to the limb is now shut off for five to ten minutes by *maintaining* a pressure exceeding the systolic pressure. (There seems no good reason for keeping the limb in the water bath during the application of the pressure, as Lewis directs, provided the room is warm, 78° F. or over.) On release of the pressure, the relaxed vessels are rapidly filled so that the skin becomes bright pink, to the very tips of the toes, in two to five seconds.

Should the blood vessels be diseased, the toes may not turn pink for half a minute or even longer. Or some toes will color long before others, giving information as to the areas most seriously deficient. The *flush* dies out most rapidly in the parts in which it first appears and lasts longest in the regions most slowly colored. An organic deficiency is clearly revealed by this test and can usually be distinguished from vascular spasm. In the latter case, the extremity, cool, damp, and tending to be cyanotic, beforehand, is made to flush like a normal limb. Or if spasm is superposed upon some degree of organic con-

by a return to a normal if only faintly pink color But in the presence of a feeble circulation a blanched elevated part regains its natural color only after many seconds The experienced observer is able to harmonize the different periods of delay in the return of color to a pressure spot (the limb being slightly elevated) with other tests

SIMPLE TESTS OF VASCULAR OBSTRUCTION AND OF CAPACITY FOR VASODILATATION

Though the more elaborate tests are required to ascertain the exact state of the arterial supply to a limb, one or two simple ones will give a surprising amount of information

Elevation and Depression —The leg having been exposed in a warm room for ten to fifteen minutes to obviate any accidental vasomotor constriction, is examined for surface temperature and color, the patient lying supine It should now be raised to an angle of 30°–45° with the body and held in that position for a period of perhaps two minutes The color of the foot is noted, that is, whether it retains a healthy pallor or takes on a cadaveric, yellowish white shade In the latter case, the arterial supply is deficient, possibly, if the blood pressure is low, only relatively deficient In this position also the characteristic cramp of intermittent limp is easily brought out automatically or by exercising the foot, and, as Samuels points out, such exercise emphasizes any deviation from the normal pink color of the sole, especially any difference between the two feet

On lowering the leg, the patient sits against the edge of the bed, the legs resting on the floor If the circulation is normal, a flush appears *in a few seconds*, first at the ankle, *and quickly* spreads to the feet and toes which take on a pink blush without blueness The whole process takes perhaps five to ten seconds or less If, however, the arterial supply is imperfect, there may be a pause of ten or more seconds before the flush appears on the foot at all, after which it usually progresses in a deliberate and perhaps irregular way, slowly reaching the

vasomotor system now in use. The next step, still a clumsy one, was the trial of various general anesthetics for a similar purpose. But, though any anesthetic capable of carrying a patient into the stage of full relaxation (and even some gases which hardly go so far) paralyzes at the same time the sympathetic nerves, such a method is hardly adapted to general use.

The solution of the problem was found in regional anesthesia by procaine. Spinal anesthesia confers a complete sympathetic paralysis upon the region anesthetized, which in the great majority of cases is the lower half of the body. It seemed at first to serve ideally for a study of the circulation in the legs, and still is the most definitely positive method of securing full vasodilatation of the lower limbs. However, when White proved that blocking the sympathetic supply to one limb could be accomplished by paravertebral injection of the various sympathetic ganglia or by anesthetizing a great nerve trunk, he opened the way for comparing the state of two symmetrical limbs—a most desirable event. It became clear also that single peripheral nerves, the sole source of sympathetic impulses to certain cutaneous fields, could be blocked with procaine, a matter explained by the much earlier anatomical observations of Kramer and Todd, Potts and others. (See Figure 2.)

The story would be incomplete without reference to the vasodilatation made possible, principally through the observations of Lewis, that heating the blood of *one* part of the body sets off a physiological reaction (vasodilatation) which warms *all* the limbs. Coller and Maddock found that heating the body raises the surface temperatures of the exposed extremities and that vascular deficiencies cause this mechanism to fail. Gibbon and Landis went a step farther and secured vasodilatation in the feet by immersing the arms in hot water.

As for drugs, a good drink of alcohol gives a very efficient dilatation, and recently Beck and De Takats have found that the administration of a standard dose of sodium nitrate offers

striction, the reactive hyperemia will be rapid, but the flushing will fall short of completeness All such fine points will only be settled by the more elaborate tests which make use of the surface temperature, or the actual blood flow through the extremity, as measures of the actual degree of vasodilatation

Tests Requiring Special Apparatus

To be accurate, tests for vasodilatation should record the actual surface temperature and insure in the field to be studied the most complete vasodilatation possible Practically speaking, there is no such thing as an accurate record of surface temperature—the radiation of heat being at the mercy of too many influences—but the use of the thermocouple is decidedly the most satisfactory method It is in securing vasodilatation that the variety of methods becomes confusing A brief review of these will explain why certain ones have survived to become standard

The test by the introduction of foreign protein, particularly typhoid vaccine, was introduced by Brown (1926) Such a substance, after causing a preliminary vasoconstriction (chill), calls forth a high fever of a few hours' duration and a universal relaxation of the blood vessels The vasodilatation and rise of temperature, according to the principles already laid down, are most marked in the digits And by balancing this rise in the tips of the extremities against that of the body temperature (*mouth or rectal*) a ratio or vasomotor index can be determined In simple language, the greater the rise of temperature upon the digits in proportion to the rise of temperature in the body at large, the more completely vasoconstriction has been relaxed and vasodilatation secured Moreover, even if no physiologic or pathologic vasoconstriction has been present, the test reveals whatever capacity sclerosed vessels have for relaxation Unfortunately, it is disagreeable to the patient, on account of the chills and malaise, and is not without danger It is *not only* variable, but both surface and body temperature are unfixed However, it opened the way to the more reliable and convenient methods of examining the peripheral

that he feels no sense of coldness or of heat. When the skin temperatures are running at a constant level, which will usually be somewhere below 80° F. (27° C.) the patient should be turned upon one side for the spinal anesthesia. In this position the contact wires will not be disturbed. He should be uncovered as little as possible.

For most adults, procaine crystals to the amount of 150 mgm. in four ccm. of spinal fluid will give a satisfactory anesthesia up to a point well above the umbilicus. The needle should be inserted into the second or third lumbar interspace. The injection is made with the patient's body horizontal, after which he is returned to the supine position. The foot of the bed may then be raised four inches, though this is hardly necessary. In any case, the head should *not* be raised on a pillow during the anesthesia (danger to medullary centers). If the blood vessels of the legs are capable of vasodilatation, the rise of temperature will be maximal, that is, to 91°–95° F. (33°–35° C.). If the vessels are in a state of spasm, such spasm will be relaxed and the same high temperatures will be reached. But if organic constriction is present—arteriosclerosis or thrombo-angiitis obliterans—the rise will be altogether absent or of only a few degrees. Expect the rise in five to ten minutes except in cases of obstinate vasospasm when it may be delayed for fifteen to twenty minutes. Any anesthesia which extends upward to the level of the clavicles will cause full vasodilatation of the arms.

Paravertebral Anesthesia.—For practical purposes, this is the only method of securing paralysis of vasoconstriction for one entire limb. If successful, the degree of vasodilatation in that limb is, theoretically at least, as complete as is the case with spinal anesthesia. Those who desire the most detailed description should consult the writings of White, who first advocated the procedure, and of Flothow, who has gone on logically to develop it into a means of securing a prolonged vasodilatation by the injection of alcohol. The accompanying illustration will bear out the statement that precise directions are of little value. The operator must have the ability to visualize the spinal column, its transverse processes and the ribs, and

a safe and reliable means of studying the capacity of th peripheral arteries for relaxation

All such procedures must be judged by the completenes with which they abolish vascular spasm In general, spina anesthesia for the lower limbs is most reliable Heating th body, or a part of it, is subject to the objection that any on vascular spasm may be so severe as to fail to be influenced Paravertebral nerve block would be ideal if it were not a littl too dependent upon the skill of the operator And periphera nerve block only affects the peripheral vessels, that is vasomotor control of the great arteries central to the block i unaffected Nevertheless, both the local application of hea and the various regional anesthesias are extraordinarily in formative—all the more so when means of taking surface temperatures are at hand

Modern means of recording surface temperature were firs developed by Benedict and his associates Now various instru ments based on the thermocouple are available None i strictly accurate The most one can say of any is that with a moderate amount of care, the changes in the temperature of *any one part can grossly be recorded and comparisons made* between two symmetrical limbs For the most accurate work a room, free from drafts, whose temperature can be controlled is required Self recording apparatus is very convenient Doubtless the technique in this field will continue to develop

Spinal Anesthesia —To take full advantage of the complete vasoconstriction secured in the anesthetic field by spinal anes thesia, cutaneous temperatures should first be taken from at least four points upon the feet, by wire loops attached with light silk about the toe or instep, during perhaps an hour's observation in a room neither hot nor cold but kept at a con stant neutral temperature—anywhere between 70° and 75° F or 21°–24° C The four points should be the dorsal surface of each great toe just behind the nail and the instep of each foot (The thickness of the skin of the sole makes this surface un reliable) While the records are being made, the thighs and body of the patient should be covered with light blankets so

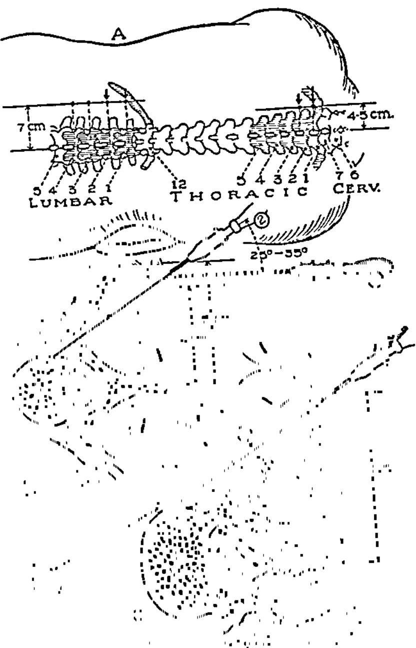


FIGURE 3. SYMPATHETIC BLOCK FOR THE ARM AND LEG For the arm, the points indicated by arrows are opposite the spinous processes; for the leg, opposite the interspaces. In making the injection of procaine, it is usually sufficient to inject at *T*, 1 for the arm and at *L*, 2 for the leg.

to know where the point of his needle is going Above all he must have the disposition suited to local anesthesia, the gentleness, deliberation and power of assurance without which injections of procaine are a trial to the patient, in some cases, an agony

Upper Thoracic Block (the Arm) —The sympathetic ganglia at this level lie about opposite the middle portion of the vertebral body, that is, from front to back, and very close to the pleura To reach them safely the needle must penetrate between two ribs, and, about an inch (two and a half cm) deeper, that is, in the direction of the front of the vertebral body, hit the body If the injection is made with the needle in this position the solution must bathe the sympathetic chain

The ganglia to be reached are the first and second thoracic Injection of the first usually paralyses the whole sympathetic supply for the arm, but injection of the second, as well, is occasionally required Wheals are made four to five cm lateral to the upper two thoracic spines and through these, using a longer needle, procaine is injected down to the sensitive surface of the underlying ribs The eleven cm needle (with a filler, and without an attached syringe) is now *thrust in directly* until it meets the posterior angle of the rib It is then withdrawn, directed toward the midline at an angle of about 25°–35° until, at a depth of about an inch (two and a half cm) frontal to the rib, it meets the side of the vertebra Here the operator's sense of position must guide him If the patient is large and heavily muscled he will have started his insertion fully two inches (five cm) lateral to the spine and will have farther to go to reach the vertebral body In any case, if he feels he is striking the body too far posteriorly he must alter the direction of his needle and push for a contact a little farther *forward*

On inserting the first needle, he will do well to attach an empty syringe and suck to see if he can draw out either air or blood If he secures air, he must have punctured pleura and lung and must reinsert the needle, hugging the vertebral column closer If blood, he must withdraw and reinsert the needle

ccm. of one per cent procaine are injected. Actually, such an injection opposite the second lumbar interspace will usually block the whole lumbar sympathetic chain (since a liberal amount of the procaine solution passes freely up and down the retroperitoneal cleavage plane) but the third interspace is often injected for additional assurance of completeness. An injection of one side will occasionally pass across the mid-line, affecting the opposite leg.

It should be remembered that any anesthesia which may result will pass toward the pubes and that the patient can experience no numbness in the leg or foot. Thus the sensation of warmth, and in case pain is abolished, of comfort, is very readily noticed. A comparison of the state of the two legs is of course a prime object of the test.

Peripheral Nerve Block.—The vasodilatation secured by an injection of procaine about a great peripheral nerve such as the posterior tibial, median, or ulnar, falls short, as Morton and Scott have proved, of being complete; that is, after a rise of surface temperature to a constant level has been obtained by the peripheral injection, spinal anesthesia will evoke a still further rise. The failure of the peripheral block to secure a maximal vasodilatation is presumably due to the normal, high vasomotor tone in the great arteries central to the block, a tone untouched by peripheral nerve block but abolished by spinal (and probably paravertebral) anesthesia. To bring out this point, one of Morton and Scott's charts, somewhat simplified, is herewith [illegible] the [illegible] a l [illegible] F. (22° C.) to at least 89° F. (31.5° C.) and as a rule nearly to 93° F. (34° C.), but posterior tibial block only raises the temperature of the great toe to between 87° F. (30.5° C.) and 89° F. (31.5° C.). In fact Morton and Scott go so far as to establish a "*normal vasodilatation level*" for both spinal (or general) anesthesia and for peripheral block anesthesia. These levels are subject to correction for variation in the room temperature (0.54° F. or 0.3° C. to be added for every degree of room temperature above 68° F. or

a little higher or lower After inserting the needle for the first thoracic (stellate) ganglion, making suction and injecting a few drops of procaine to see if the patient tastes the fluid or coughs, five to twenty ccm can safely be introduced

A successful block of the stellate ganglion will cause contraction of the pupil as well as dilatation of the peripheral vessels of the limb (if these are dilatable) A subjective sense of warmth will be felt in the hand, whose surface will become dry and warm to the touch The cutaneous temperatures should reach a peak in a few minutes By inserting the needle from *above the first rib—in a direction more cauded than usual*—the risk of piercing the pleura and lung is less and the pupillary reaction is an even surer test of success

Lumbar Block (the Leg)—For the lumbar injection the technique must be varied slightly on account of the greater size of the vertebral body and of the forward position of the sympathetic chain The wheal is placed opposite the interspace instead of the spinous process and at a distance of about two and a half to three inches (six to seven cm) from the midline To fall opposite the second lumbar interspace it must be made very close to the twelfth rib Through the wheal opposite the second lumbar interspace, the muscular aponeurosis, which is sensitive, is carefully infiltrated with the one per cent solution The long needle often meets the tip of a transverse process and helps in the estimation of depth, for the point to be reached is rather over an inch (three cm) anterior to the process and more than three inches (eight cm) from the surface The needle is thrust in at an angle of about 45° toward the vertebral body, just meeting the body full on its lateral aspect The succeeding adjustment requires some skill The needle is first withdrawn and redirected farther forward striking the body at a still greater depth Then, ideally, it is adjusted so that it glides past its last contact with the body to a depth one fourth to one half an inch (one cm) greater Here its point lies on the anterior edge of the psoas muscle and just behind the aorta or vena cava, here suction is made to determine whether or not a blood vessel has been injured and here ten to twenty

upper edge of the great trochanter to the posterior superior spine (iliotrochanteric line), a perpendicular is drawn downward upon which a point is selected one and a quarter inches (three cm.) from the first line. Here a needle, four inches (ten cm.) long, is carefully inserted in a direction normal to the surface until it causes paresthesia in the course of the sciatic or meets bone at a depth of two and one-fourth to three inches (six to eight cm.). The nerve should be looked for at about a depth of two inches (five cm.). It may be necessary to feel for the nerve which should not be pierced. Ten to twenty ccm. of a two per cent solution of procaine may be injected. Anesthesia should appear in ten to twenty minutes. It is doubtful whether sciatic block can be expected to give a maximal vasodilatation.

Posterior Tibial Block.—This block gives, for normal vessels, an incomplete but constant vasodilatation and rise of temperature to between 87° and 89° F. (30.5° to 31.5° C.). The nerve is found below the internal malleolus, where it is easily palpated as a hard round cord behind the posterior tibial artery. Having made a wheal over it with a fine needle, fix the nerve with the index finger of the other hand and introduce ten ccm. or so of two per cent procaine into the fascial compartment in which the nerve lies. The nerve itself should not be pierced. The skin of the heel, sole, and plantar surface of the toes soon becomes wholly or partly anesthetic and a flush appears. The skin of the sole is so thick that the temperatures are best taken from the plantar surfaces of the outer phalanges of the great and little toes.

Median Nerve Block.—Though the median can be blocked at the wrist, its large size and tough sheath make its injection rather unsatisfactory as a means of securing a peripheral vasodilatation. The guide is the tendon of the palmaris longus muscle. By resisted flexion of the wrist this tendon and that of the flexor carpi radialis (on the radial side of the former) are brought out. The nerve lies between the two. Through a wheal at the level of the radial styloid, just lateral to the tendon of the palmaris longus, the needle is introduced directly through the deep fascia. When the vicinity of the nerve is reached,

20° C) but for practical purposes are those described above The usefulness of peripheral block anesthesia lies, of course, in the fact that the temperature level it secures is just as reliable a test of peripheral vasodilatation as spinal or general anesthesia The defect of the method really lies in the failure of procaine in some cases fully to anesthetize a great nerve such as the sciatic, posterior tibial, median, or ulnar

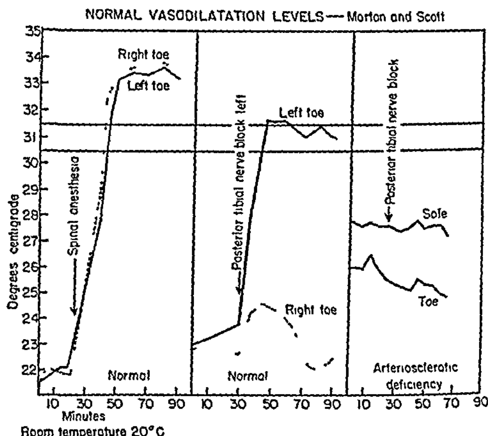


FIGURE 4 NORMAL VASODILATATION LEVELS FOR SPINAL AND PERIPHERAL SYMPATHETIC BLOCK (after the Charts of Morton and Scott)

The Sciatic Nerve—The nerve is reached as it comes out through the great sacrosciatic foramen, just below the pyri formus muscle Labat's technique is the following

The patient lies on the side opposite to the one to be injected in the Sims position, that is, both legs drawn up, the knee corresponding to the nerve to be injected a little overlapping the other From the middle of a line traced from the

controlled room, especially when means of continuously recording surface temperature are available, affords perhaps the most luxurious and foolproof means of studying the vasomotor reactions in the extremities. The temperature of the digits having been recorded in a cool room, the body can be heated to secure the maximal vasodilatation.

Tests by the Use of Drugs

Alcohol.—A stiff drink of whiskey or any strong liquor is, except for the corruption of the individual, an admirably simple and practical method of testing the ability of the vascular bed to dilate. Unless a very accurate check on such vasodilatation is needed, the oscillometer will give a sufficiently good idea of the patient's response. His own sensations will usually inform him that the dose has been sufficient and that all the dilatation possible under the circumstances has been secured. The test is of some value in estimating very *roughly*, in the presence of an intermittent limp, the elasticity of the circulation and the possibility of improving the arterial supply to a limb by (permanent) sympathetic block. Some individuals, for instance, will be able to walk *twice as far*, without limping, after the d[illegible] d

[illegible] *of sodium nitrate*, intravenously, in a dose of one ccm. of a four per cent solution has recently been advised by Beck and De Takats. The oscillometer is used to record the amount of vascular relaxation. The effect appears in ten to fifteen minutes and lasts perhaps an hour. In this dosage, the drug apparently is not dangerous.

Arteriography

By exposing a limb to the X ray during the injection of certain solutions * opaque to the X ray, much information has been secured as to the exact state of the arteries in vascular

* Thorium dioxide, or "Thorotrast" has been [illegible] substance [illegible] ably [illegible]

paresthesia in the median field is apt to be noticed Ten to twenty ccm of a two per cent solution of procaine is then introduced If no paresthesia appears, the needle can be directed a little farther radialward Massage aids in bringing on the anesthesia

Ulnar Nerve Block—This is best made at the elbow where the nerve is easily palpable posterior to the internal epicondyle of the humerus Through a wheal over the nerve a fine needle is introduced into its vicinity (not actually piercing it) A two per cent solution of procaine is liberally injected The area supplied by the ulnar nerve becomes pink and dry, giving the patient, as a rule, a sensation of warmth and numbness, the anesthesia being incomplete but vasodilatation satisfactory

Exposure to High and Low Temperatures

The most practical means of using heat to secure vasodilatation is to immerse the hands and arms in hot water The usual arm basins serve the purpose The water should have a temperature up to 110° F (43° C), that is, a heat just bearable If an accurate account of the rise of surface temperature is to be kept, the usual preliminary control observations should be made at a room temperature of about 70°–74° F An immersion of fifteen minutes will usually cause a rise of temperature in the feet, and, except in the face of an obstinate vasoconstriction, the rise will in most cases be maximal Obviously *the test can be reversed Beginning with warm surroundings,* and a high level of surface temperature for the feet, the arms can be immersed in cold water But here the fall will merely record the promptness of the resulting vasoconstriction There is no ideal end point for the observation On the whole, the responses to immersion of the arms in hot water can not be given full authority Lack of response in the form of a rise in the surface temperature of the feet can not be regarded as proof that no possibility of vasodilatation exists To test the arms, the feet and legs can be immersed in hot water, a rather awkward procedure and not entirely satisfactory

A jacket, electrically heated, for use in a temperature

controlled room, especially when means of continuously recording surface temperature are available, affords perhaps the most luxurious and foolproof means of studying the vasomotor reactions in the extremities. The temperature of the digits having been recorded in a cool room, the body can be heated to secure the maximal vasodilatation.

Tests by the Use of Drugs

Alcohol.—A stiff drink of whiskey or any strong liquor is, except for the corruption of the individual, an admirably simple and practical method of testing the ability of the vascular bed to dilate. Unless a very accurate check on such vasodilatation is needed, the oscillometer will give a sufficiently good idea of the patient's response. His own sensations will usually inform him that the dose has been sufficient and that all the dilatation possible under the circumstances has been secured. The test is of some value in estimating very roughly, in the presence of an intermittent limp, the elasticity of the circulation and the possibility of improving the arterial *supply to a limb* by (permanent) sympathetic block. Some individuals, for instance, will be able to walk twice as far, without limping, after the dose of alcohol as before. On the whole, the method is rather on the rough-and-ready side—for a snap-diagnosis.

The use of sodium nitrate, intravenously, in a dose of one ccm. of a four per cent solution has recently been advised by Beck and De Takats. The oscillometer is used to record the amount of vascular relaxation. The effect appears in ten to fifteen minutes and lasts perhaps an hour. In this dosage, the drug apparently is not dangerous.

Arteriography

By exposing a limb to the X ray during the injection of certain solutions * *opaque to the* X ray, much information has been secured as to the exact state of the arteries in vascular

* Thorium dioxide, or "Thorotrast" has been [illegible]
substance [illegible]
ably [illegible]

disease of the limbs The variations in the caliber of arterio-sclerotic vessels, the situation of an obstruction in the form of an embolus or thrombus, and above all the nature of the collateral circulation in serious arterial deficiencies have been observed Yet it cannot be said that arteriography is of any vital diagnostic aid, and perhaps so far it has been most successful in confirming the impressions acquired by simpler tests It should probably be practiced only by the most expert, not entirely because of the danger inherent in the procedure but *because perfect technique alone justifies an exact interpretation of a picture in a critical case*

There are minor variations in the method of introducing the opaque material, depending upon whether the act is performed by a "team" or by an individual There is, however, a general agreement upon the following points

1 To visualize the vessels of the forearm and hand the injection is made into the brachial artery just above the elbow At this level five ccm of the solution are sufficient and more than ten ccm are never required

2 The vessels of the forearm and hand are best visualized in the antero posterior position

3 To visualize the vessels of the lower thigh, the knee and the leg, the injection is made into the femoral artery within the femoral triangle (below the giving off of the profunda) The best view is secured when the exposure is almost lateral, the leg externally rotated and the knee placed against the plate Twenty ccm of the solution are required

4 To visualize primarily the vessels of the leg, ankle and foot, the injection is made into the femoral artery in Hunter's canal The exposure should be made in an antero posterior direction Not more than twelve ccm of the solution are required

The method used by Veal and McFetridge for injection into the femoral artery is the following The skin over the site of

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serious damage Such damage however has not yet been proved inevitable in the dosage used

Iodine solutions, such as are used *for the* purpose of making intravenous pyelograms, give less vivid shadows but are without danger to the individual Some of them however cause pain when injected Such solutions as "Diodrast" seem reasonably satisfactory Doubtless new and superior ones will be invented Thus far, all the solutions used cause some degree of vascular spasm.

injection is anesthetized with one per cent procaine. The artery is punctured with a number eighteen needle attached to a syringe filled with the solution. As soon as the puncture is accomplished (bright red blood enters the syringe in spurts) pressure is made with the thumb just proximal to the site of the puncture, stopping the spurt of blood into the syringe. At once, the injection is made, the thumb continuing to compress the artery. When all the solution has entered the vessel, the pressure of the thumb is released, allowing the distal tree to be filled. Three to six seconds later, depending upon the position of the puncture and the length of the limb, the exposure is made.

For the brachial injection (at the elbow) a Wassermann needle is used, the syringe containing the solution being attached. A local anesthetic is hardly required. Immediately upon entry into the vessel, digital pressure is made just proximal to the puncture. Pressure is released and the exposure is made as soon as the injection is concluded.

The method of injection used by Allen and Camp is somewhat different. For the brachial injection, a blood pressure cuff is first placed upon the upper arm as near as possible to the shoulder. The artery is punctured with the needle attached to the syringe containing the solution. When blood spurts back through the needle, the cuff is inflated to systolic pressure, shutting off the circulation. The solution is then injected, the needle withdrawn and an exposure is made at once. But now the cuff is deflated to the diastolic level for two to four pulse beats. Upon its re-inflation, to shut off the flow, a second plate is taken. The procedure can be repeated for the taking of a third plate. Obviously this procedure requires nice team work.

For the femoral injection, the needle is introduced, the artery shut off proximally by pressure with the fingers, and the injection made. On withdrawing the needle, the artery is released for a few beats, an exposure made, and the vessel again compressed. As in the case of the arm, the release, exposure, and compression can several times be repeated.

Blood Flow as a Test of the Arterial Circulation

The flow of blood through the foot has recently been used by Kunkel and Stead as a measure of the efficiency of the circulation in the lower extremity In their publication, they state that they have modified for this purpose the apparatus of Hewlett and Van Zwaluwenburg as well as that of Freeman, intended to measure the flow of blood in the hand The apparatus is a plethysmograph (water bath) in which the foot is first accustomed for half an hour to a temperature of 33° C "When the venous outflow is occluded by a 'collecting pressure' lower than the diastolic pressure, the rate of the initial increase in the foot volume is a measure of the amount of blood flowing to the foot" The result is expressed in so many ccm per minute per 100 ccm of foot

Expressed as above, the average blood flow of normal subjects was found to be 17 1 ccm (the highest 25 9, the lowest 11 1 ccm) This is about one half the flow estimated in a similar way for the hand The investigators found that a fifty per cent reduction in the flow of arteriosclerotics and sufferers from thrombo angiitis obliterans was not associated with symptoms or external signs Beyond this point, evidences of arterial deficiency were usually noticed A rather interesting finding was the discovery that an intermittent limp might be present though the flow was seemingly sufficient to have obviated it That is, the muscles of the calf might be ill supplied, though the foot received a good flow of blood

The test is interesting as a check on others but seems, at the moment, to hold no advantage over less cumbersome methods However, the apparatus, when perfected, should be far less expensive than an oscillometer or the thermocouple galvanometer machine for recording surface temperatures

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CHAPTER II

ARTERIOSCLEROTIC DEFICIENCY AND THROMBOSIS

The pathological background of arteriosclerotic peripheral vascular disease is narrowing of the vascular channel due to a chronic, progressive thickening of the intima This is not an orderly or an evenly distributed process There is intimal proliferation, generally most marked on one side or another of a vessel, so that a semilunar thickening, as seen in cross section, is built out into the stream Hyperplasia of the elastic tissue in such areas is followed by atheroma and calcareous deposits By such means the lumen is narrowed and elasticity is lost Just what makes this process vary so much from individual to individual is a mystery So far as the limbs are concerned, it is always more advanced in the lower than the upper, perhaps because arterial pressures are decidedly higher in the legs than the arms One can adduce as causes mental and physical strains, infections and other influences over which human beings have little control, but of all adverse factors diabetes seems to be the most powerful Diabetics suffer at an earlier age than do others from arterial deficiency in the legs, and apparently insulin does not protect against this change

Arteriosclerotic narrowing and hardening leave a vessel of irregular caliber Fibrous thickening and deposits of calcium are most marked at bifurcations and points of active bending Thus the vessels of the groin, the popliteal region and upper calf in particular are most seriously affected Once encroachment on the lumen has begun, there is a tendency to a deposit of platelets and so to thrombosis A sudden closure brought on in this way cuts off the arterial stream from a considerable area and causes ill nourishment, if not actual gangrene, in the

field served by the occluded vessel. By contrast, gradual constriction of an artery is accompanied by the development of so effective a collateral circulation that the peripheral parts may remain well nourished. On the one hand, anoxemia is sudden and gangrene follows: on the other, the opening of new channels so nearly keeps pace with contraction of the old that function need never be disturbed. When the development of a collateral circulation barely keeps pace with arteriosclerosis, the stage is set for intermittent claudication and the so-called "trophic" disorders of the nails and skin, a state of things which may be prolonged, without any *serious* disability, for many years.

In arteriosclerotic deficiency, the small collateral arteries, *like the* main vessels, are often irregular in caliber and so erratic in distribution as to permit a more satisfactory blood supply to reach one area than another. The toes, perhaps, receive a sufficient circulation but the muscles or some one group of them does not. Or the muscles are well looked after and some or all of the toes are ill-nourished. Naturally the great muscles, which require *far more blood* in action than at rest, are unable to function normally. The individual finds that after walking several blocks, one leg *or the other* feels numb or cramped, or even as if stuck with a knife. He rests for a minute or two, finds himself comfortable and steps out again, only to have the same pain return after about the same distance is covered at the same pace. This intermittent limp may become worse or remain fixed at the same point or may improve, according as the collateral circulation is able to respond to the deficiency. But it is important that its nature be recognized, and a misfortune when the patient's disability is attributed to "fallen arches".

In a recent study of arteriography, Veal and McFetridge show how the pathological background can explain the clinical appearances; for example, that a relative lack of blood vessels among the great muscles of the leg corresponds in most cases to the clinical signs of arterial deficiency, that is, to the severity of an intermittent limp. By contrast with the abundant net-

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Arteriosclerotic narrowing and hardening leave a vessel of irregular caliber. Fibrous thickening and deposits of calcium are most marked at bifurcations and points of active bending. Thus the vessels of the groin, the popliteal region and upper calf in particular are most seriously affected. *Once encroachment on the lumen has begun, there is a tendency to a deposit of platelets and so to thrombosis.* A sudden closure brought on in this way cuts off the arterial stream from a considerable area and causes ill nourishment, if not actual gangrene, in the

or female, perhaps more often a male, but thrombo-angiitis obliterans in a female is almost unknown. An arteriosclerotic deficiency is either so well balanced by a collateral circulation as to cause only minor disorders to which the individual gives little attention or it develops rather rapidly into a serious local or extensive gangrene. Perhaps only one individual among ten who show minor signs of arteriosclerotic deficiency ever comes to ulceration or gangrene, but that one may suffer from an extensive necrosis of the toes or a foot after a very short period of premonitory cyanosis and pain. By contrast, the pregangrenous stage of Buerger's disease may be prolonged for years, and its actual gangrene need never be very extensive, which is another way of saying that symptoms of circulatory deficiency threatening ulceration or gangrene are seldom noticed in arteriosclerotic disease for more than a few months, but in thrombo-angiitis obliterans may be present for years. Again, though arteriosclerotic deficiency, as between the legs and arms, shows itself almost exclusively in the legs, there are often telltale signs of arteriosclerosis elsewhere, notably dizziness, transient aphasia, lack of tolerance for cold, and the arteriosclerotic vessels appear calcified to the X ray. By contrast, the signs of thrombo-angiitis obliterans are noticeable chiefly in the legs or only very late, in the arms, and calcification of the arteries is rare and much delayed. In earlier times, perhaps before cigarette smoking became general, it seemed that thrombo-angiitis was confined to Polish or Russian Jews. And though such is no longer true, the disease, in the United States at least, is seen more often in Hebrews than in any other race. Finally, arteriosclerotics never suffer from superficial "wandering" phlebitis, whereas some observers have maintained that phlebitis migrans attacks those suffering from Buerger's disease in as much as thirty per cent of all cases.

The Presenting Symptoms.—Intermittent limp, if the story could be dragged out of every patient, would probably be found to be the most common initial symptom of arteriosclerotic deficiency. But among those who present themselves for

work of very fine arteries and arterioles so often seen in thrombo-angiitis *obliterans*, *the individual arterial* branches are fewer and more irregular in caliber In both diseases, the total arterial circulation may be equally lacking and the functional difficulty much the same, but in arteriosclerosis the way is laid for *ultimate gangrene on a larger scale* The clinical application of these characteristic pathologic changes will again be discussed when the course of the two diseases is compared

There is a pathological variation upon arteriosclerotic deficiency known as *Monckeberg's arteriosclerosis* This is characterized by sclerosis of the media rather than the intima, is thought to occur a little earlier in life than the common form, *and is not so apt to exhibit calcification to the* X ray However, it may not be an independent process but rather a stage of the usual disease, and that it can be distinguished as a clinical entity is exceedingly doubtful Its supposed peculiarities will be considered with the clinical manifestations of arteriosclerotic deficiency and in particular with the differential diagnosis between this state and thrombo angiitis obliterans

Arteriosclerosis and thrombo angiitis obliterans include ninety-five per cent of all arterial deficiencies Of this percentage arteriosclerosis has much the larger share Both diseases produce their effects by arterial narrowing or obstruction, so that they necessarily have a family resemblance It is in their background and in the progression of their signs and symptoms that the two diseases mainly differ And though it may happen that at any one moment in the course of each, and in the unusual individual whose age, sex, and race are consistent with either disease, a differential diagnosis is difficult, the etiological factors contrasted in the following paragraph will greatly aid in making a distinction between them

The victim of arteriosclerotic disease is rarely under fifty years of age, the average of those who first complain of symptoms being perhaps sixty, in contrast with thrombo angiitis obliterans which usually shows itself between the ages of twenty and forty The arteriosclerotic may be either a male

haps the forefoot become red or cyanotic in the dependent position and that some particular toe is discolored before the others. He may notice also swelling, which is the next step in the self-revelation of the deficient circulation, but these are rather objective signs to be noted by the observer.

The Clinical Signs of arteriosclerotic vascular deficiency, in the absence of acute arterial thrombosis or actual gangrene, again are suggestive rather than striking. Especially in the very elderly, there is almost always some atrophy of the legs, most obvious in, but not confined to, the calf. The surface of the leg, foot and toes is dry and cool. The skin is often rather thin and tending to be transparent. Such a state is, of course, consistent with a deficiency of long duration and is seldom a cause of complaint. But when gangrene is imminent or is actually present, the natural color of the skin will usually be changed. Then some one or several of the toes may be red or even bluish red. The discoloration may run up upon the foot itself. By contrast with thrombo-angiitis obliterans, such redness or cyanosis is less common and when present, is less well marked. Edema of the toes and forefoot is likewise less important in arteriosclerotic deficiency than in Buerger's disease —a difference of degree only and not a reliable distinction. Both cyanosis and edema are likely to come out best when the foot is left dependent for considerable periods. Should spontaneous pain be present it will likewise be increased by letting the leg hang.

Minor Gangrene, in the absence of diabetes, which will be discussed in a separate section, is usually well localized. In the type which follows a slow atrophying sort of arterial occlusion and in which a gross occluding thrombosis is not a factor, it will often have begun beside the great toe-nail, as already described, involving part of the outer phalanx, but will not reach the base of the toe. Or if one of the middle toes is particularly deformed or calloused, a local ulcer associated with very little gangrene may have formed. A callus over the prominent metatarsal head at the base of the great toe is a common site of gangrene and a corn upon the little toe is

the treatment of serious pain, ulceration, or actual gangrene, it is not the usual presenting symptom, which actually is numbness and coldness of one or both feet Very likely, the state of the arteries has long prevented the individual from walking any considerable distance or at anything more than the slowest pace Yet having been broken slowly to this situation, he, or she, seldom thinks to complain of the cramp like *numbness, excited by locomotion, which disappears so quickly* on rest See many an elderly woman on a street crossing It is not necessarily stiff joints which makes her move so slowly She just can't walk faster and knows it Only if she hurried would she exhibit the limp of arterial deficiency Intermittent limp, already touched upon, will be more fully described in the chapter devoted to thrombo angiitis obliterans

Beside the characteristic feeling of coldness, of tingling, and of a cottony sort of numbness, there is sometimes a localized pain in the foot principally affected, a pain which comes and goes, at first, without much reason but which tends to become constant It may be felt in the sole, the instep, the heel, or in the toes Such a pain or a feeling of coldness of the feet may prevent sleep at night, but even if such troubles are absent, night cramps frequently occur These are apt to be confined to the foot or to some particular group of muscles of the leg, often causing violent dorsiflexion of the great toe or all the toes

Trophic changes are common, cracks, especially upon the *heel, and thickening and deformity of the nails* This last may result in unskillful cutting of the nails or of attempts to treat what seems to be, but probably is not, an ingrowing toenail upon the great toe An individual often complains of soreness from the irritation of a deformed nail and ends by developing gangrene of the great toe as a result of the trauma and infection brought on by an ill advised attempt at a surgical cure Indeed, treatment of the corn, callus, or nail with the knife is very commonly a beginning of the familiar series of events, infection, ulceration, and gangrene

The patient is occasionally aware that the toes and per-



ARTERIOSCLEROTIC GANGRENE. *A.* R. K. G., a mild diabetic, sixty-eight years of age, symptoms of only a few weeks' duration, local gangrene, absent peripheral pulses, calcified vessels. The slough was finally cast off, with healing. *B.* J. J. B., sixty years of age, symptoms of one week's duration, local gangrene beside deformed nail, absent peripheral pulses, calcified vessels. Recovery under local treatment of nail and wet dressings. *C.* Moist Type due to thrombosis. P. N., symptoms for two years; cyanosis and numbness for three weeks, demarcation beginning.

PLATE I

especially likely to touch off an ulcerative, gangrenous process With all such small lesions there will sometimes be found necrosis of a phalanx, a process slowly suppurative and detectable by the X ray but hardly worthy of the name of osteomyelitis A sinus may lead down to such a spot or to an infected joint, but as compared with similar states in diabetics, there is little redness and tenderness, that is, external evidence of infection Tendons and tendon sheaths are relatively seldom involved

Any area of gangrene, if seen from the beginning, will first appear a deep purple, turning slowly to black Not for a week or more will the border of such an area become clearly defined, but as demarcation proceeds, the gangrenous part will shrink and dry Meanwhile, the adjacent skin is becoming a little red and swollen The point is often made that when the outer half or all of a toe is gangrenous, the process cuts just as definitely through the deeper part as through the skin and should be allowed to take its own pace, so that final separation, which only takes place after several months, is given the opportunity to leave a granulating surface capable of healing The proximal zone of reactive hyperemia and induration is in the meantime a protection against infection To amputate the gangrenous toe of arteriosclerosis is very hazardous and only those thoroughly familiar with the disease should ever attempt it Nature's method is slow, to be sure, but safe

There is one rather notable distinction between the gangrene of arteriosclerosis and of thrombo angiitis obliterans Pain in established arteriosclerotic gangrene can seldom compare in severity with that of Buerger's disease There is a burning, agonizing quality, sometimes a feeling as if the toes were being crushed, and an associated hypersensitiveness in the gangrene of Buerger's disease which is very rare in the purely arteriosclerotic states Often the gangrene of arteriosclerosis will be sore rather than painful, though elevation and occasionally hanging the foot down may bring on considerable pain Absence of pain is even more marked in diabetic gangrene as will presently appear In such, the gan



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grenous, perhaps infected, part is often actually anesthetic. An interphalangeal joint, for instance, so far destroyed as to grate on motion, will often cause no pain whatever.

One form of senile arterial disease, undoubtedly arteriosclerotic, behaves so much like thrombo-angiitis obliterans as to be easily confused with it. The patient is in the forties or fifties rather than the sixties, that is, prematurely aged. The threatening prodromal symptoms develop more slowly than is usual in arteriosclerosis though rather too rapidly for thrombo-angiitis obliterans. The cyanosis of the toes and forefoot, or it may be the actual gangrene, is particularly painful and very resistant to treatment. Apparently a collateral circulation is not readily developed. In such cases, the arteries of amputated limbs have so often shown the medial type of sclerosis characteristic of Mönckeberg's arteriosclerosis as to suggest the existence of a clinical-pathological entity. However the matter [illegible] fer to say [illegible] thrombo-angiitis obliterans but the patient is rather too old for that disease. After all, arterial narrowing is arterial narrowing, and Buerger's disease is chiefly distinguished by its inflammatory background, the involvement of veins, and especially the youth of the subject, which permits collaterals to be opened while the arterial circulation is still elastic. There is no reason why there should not be borderline cases.

Major gangrene, often of great extent, occurs particularly when the patient is driven to seek help by *thrombosis* which suddenly closes a good sized artery, previously patent. Here the duration of symptoms preceding the actual onset of gangrene is especially short. The patient is usually struck down by severe pain, the pain of sudden ischemia, much like that of an arterial embolism. That this varies in severity from a feeling of numb coldness to real agony is well known. In many such cases the whole forefoot, or the whole foot, or the foot and part of the leg will turn pallid, bluish, and finally purplish, the color being rather splotchy and with no clear-cut limit above. The skin over such an area will be cold; the tis-

sues swollen and perhaps blistered These signs often, though by no means necessarily, lead to a moist gangrene In any case, there is a distinct difference between this picture and that of the slower mummifying process Given infection and edema, a moist type of gangrene occurs Yet there are many intermediate stages, making a distinction between "dry" and "moist" gangrene impracticable

Diagnosis—The identification of an arteriosclerotic deficiency or actual gangrene rests upon the age of the individual, an appearance of atrophy, the state of the skin, including its coolness, and the evidence of restricted circulation revealed by the simple tests outlined in the preceding chapter That is to say, it can be shown that the leg and foot are not only cool to the touch but are very slow to become warm when exposed to a heated environment and cannot be made to exhibit anything more than the slightest reactive hyperemia by the various methods described These, of course, are qualities consistent with an arterial deficiency of any sort, but in addition, the patient, who will seldom be under sixty years of age and usually a good deal older, will rarely complain of having suffered from coldness, numbness, or an intermittent limp for more than six months, and if actual gangrene is present, for a far shorter time, whereas the much younger individual suffering from thrombo angiitis obliterans will almost necessarily have noticed an intermittent limp for years The physical signs are confirmatory The toes and foot seldom become as cadaveric on elevation or as cyanotic on depression as in the correspondingly serious case of Buerger's disease Moreover, the larger gangrenes involving the foot and part of the leg and due to a rapidly developing thrombosis are far more extensive than are ever occasioned by the latter condition

Pulsation in the larger arteries often reveals a difference between the two legs but such pulsations are not to be counted on to distinguish arteriosclerosis from other deficient states A pulse in the dorsalis pedis or posterior tibial artery is fairly often present in the face of a decided intermittent limp, signs of coldness and atrophy, and even actual gangrene But all

four arterial pulsations are never palpable. Both posterior tibials will be absent and the dorsalis pedis of the leg most seriously affected will perhaps be feeble. Or all pulsations will be missing on the worst side and only barely detectable on the other. This is in contrast with thrombo-angiitis obliterans where peripheral pulsations are almost always absent once clinical signs of the disease have appeared. Sometimes in the presence of diabetic gangrene, which after all is merely arteriosclerosis with or without infection, the pulsations of several peripheral arteries are remarkably vigorous. In other words, arteriosclerotic deficiency is a patchy one, the supply to some parts being good and to others poor.

Methods of Recording and Classification.—By this time, sufficient information has been presented, in this and the preceding chapter, upon which to base a method of study and system of record. And though arterial deficiency has chiefly been discussed, the inevitable comparison with thrombo-angiitis obliterans will have given at least a partial account of that disease. Accordingly, a table of differential diagnosis is presented below and, following this, a scheme for making special notes in vascular disease. Both of these are taken in great part from A. W. Allen's publications which in turn are based upon procedures devised in the Vascular Clinic of the Massachusetts General Hospital. The writer has taken some liberties with both the table and method of notation. Mönckeberg's Arteriosclerosis has been omitted, on the ground that it is hardly a clinical entity. Vasomotor disorders, as a matter of contrast, are included though, strictly, they are too little of one type to be covered in this way. It should be realized that the etiologic, pathologic and clinical signs recorded often represent averages and tendencies rather than fully reliable indications of disease.

Special Notes

The first seven headings relate to the history. Headings 8 to 10 relate to diabetic gangrene. The final group of six relate of course to special examinations and are not equally required

TABLE OF DIAGNOSTIC INDICATIONS

	Arteriosclerosis	Thrombo-angiitis Obliterans	Vasospasm—Raynaud's and Permanent Functional Types
Age	60+	20–40	15–30
Sex	Males and females	Males	Females
Nationality	All	Hebrews 40% All but negroes	All
Duration of Symptoms	Months	Years	Years
Extremity	Lower	Lower (until very late)	Raynaud's upper Permanent type lower
Symmetry	Unilateral or one side at a time	One side at a time	Bilateral
Ulceration and Gangrene	Early after declared symptoms	Late	Late and moderate
Vessels in X ray	Calcified	Not calcified	Not calcified
Peripheral pulses	*Poor or none*	*None*	Normal
Procaine Block	No vasodilatation	Slight vasodilatation	Vasodilatation
Collateral Circulation	Present but erratic	Many fine vessels	Not present or required
Pathological State of Vessels	Thick irregular intima, atheroma and calcification	Inflammation, organized thrombosis—arteries and veins	In Raynaud's disease only, then late sclerosis of digital arteries

for all cases. They are fully described in the previous chapter.

1. The duration of symptoms: intermittent limp, coldness, numbness, trophic changes. If ulceration or gangrene is present, its preceding and prodromal symptoms: the duration of ulceration or gangrene.

2. Date and character of injury or local treatment as an exciting factor in the development of ulceration or gangrene.

3. Occupation: when disability began; how far the individual depends upon the use of the affected limb or limbs.

4. Nature and severity of pain: circumstances of its onset, its development. Is it spontaneous or dependent on the use of the limb?

5. Use of tobacco: reaction to other factors such as cold.

6. Description of the lesion: the present state of ulceration, gangrene and infection.

7. Nature of the peripheral pulsations: their presence or absence in the dorsalis pedis, posterior tibial, popliteal, femoral arteries. These to be described as + to ++++.

For diabetics: all held to be emergencies

8. Blood sugar to be studied.

9. Blood cultures to be taken if infection is present or suspected.

10. Cultures taken from all open wounds.

Tests to be recorded

11. Effect of elevating and depressing the leg: time required for blanching on elevation, and flush and cyanosis on depression (routine).

12. Reactive hyperemia in response to vasoconstriction. Length of time required and completeness as observed in toes (routine ?).

13. Surface temperatures as noted by contact with back of fingers (routine): as recorded by thermocouple; as a reaction to procaine block; peripheral; spinal (optional).

14. Oscillometry. Oscillations in the calf and their changes in response to a reactive hyperemia by any method (optional).

15 Roentgenological study of blood vessels (routine)

16 Photographs (optional)

It will be observed that except for the X ray and the bacteriological investigation, both of which are available in any well equipped hospital, all the important routine observations require no special apparatus Such are described in Chapter I, and their relative importance is there discussed

Treatment in the Absence of Ulceration and Gangrene—For the individual who complains of a deficient circulation *without gangrene—numbness or coldness, intermittent limp*, perhaps discoloration of the toes, nocturnal cramps—there is a useful routine which can be expected to lessen the patient's discomforts, retard his down hill course, and, in the younger and more elastic group, lead to actual improvement This depends upon the development of collateral vessels to compensate for the gradual closure of one or more of the larger peripheral arteries—the anterior and posterior tibial and the peroneal It should be comforting to perhaps 90 per cent of such persons to know that by giving up tobacco, curtailing their physical activities, and encouraging their circulation by vascular exercises they will be able to live in comfort and avoid the ulceration or gangrene which may have seemed to threaten them

Tobacco, if the patient is a smoker, must be barred It has nearly as unfavorable an effect upon arteriosclerotics as upon those suffering from thrombo angiitis obliterans It is especially to be avoided in those having pain, whether brought on by exercise or of a spontaneous sort And since smoking definitely causes vasoconstriction, it may be expected to interfere with the widening of the vascular bed and the development of a collateral circulation, the first objects of treatment

Fatigue should be relieved This may mean rest in bed for the tired housewife or a vacation for the business man The laborer may have to devote his days off to reclining or actually going to bed, the feet being elevated to the position of greatest comfort, which may mean the horizontal or a little below it For the indoors person, the stimulation of outdoor air and

such mild exercise as can be taken without exciting discomfort is a definite advantage. Though many individuals who complain of a deficient circulation have a high blood pressure, some have a low one, in which case the delivery of blood through sclerosed arteries will be aided by any treatment which increases the force of the heart beat. Hence proper rest, gentle exercise, and an outdoor life will be doubly effective.

The Routine Care of the Feet.—This is of great importance in arteriosclerotic deficiency and becomes even more vital in the presence of diabetes. After a daily bath in warm water and careful drying, olive oil or lanolin should be rubbed into the skin, particularly where cracks are present, as upon the toes or heels. A search should be made for scaling or blistering suggestive of epidermophytosis which may become a point of entry for pyogenic infection, and so lead to ulceration and gangrene. If the fungus is found, its treatment is a problem. Usually an ointment, such as half-strength Whitfield's, with thymol 1.5 per cent, is helpful but some patients can be treated successfully only with liquid fungicides, as for instance, 1 to 2000-3000 permanganate solution, five per cent aluminum acetate, or one to two per cent salicylic acid in fifty per cent alcohol.

The toe-nails, unless they are grossly thickened and deformed, should be cut squarely and rather long. They should always be soaked before cutting and if curved and thickened may be filed. Corns, which are so often a starting point for gangrene, should be pared down by someone expert in that line. Like the nails, they must first be softened by soap and water. Calluses should be reduced with sandpaper.

The matter of a covering for the legs is most important. Wool for socks or stockings is best. If the surroundings cannot be made sufficiently warm, lamb's wool casings for the legs at night preserve the natural heat as well as anything. Bed socks at least should be worn on cold nights. Never letting the feet be exposed to cold or actually feel cold is a constructive step in developing a collateral circulation.

Postural exercises, which will be more fully described in

the treatment of thrombo angiitis obliterans, are very well worth carrying out in arteriosclerotic states They are especially useful when discoloration of the toes and signs of edema indicate that the compensatory circulation is insufficient The physician must work out with the patient the proper routine It will be found perhaps that the legs blanch in a minute and a half when elevated to an angle of 30° (45° might be too high) and that a flush is slow to appear when the legs are allowed to hang, but that perhaps two minutes of depression will secure the maximum pink flush without cyanosis During the period of depression the feet should be exercised as directed by Allen (page 95) Finally, the legs should be wrapped in warmed blankets and kept horizontal for about five minutes The physician must devise an inclined plane for elevation, must discover how many times the cycle shall be repeated without too much fatigue and how many times a day a set of individual cycles shall be carried out

A hot sitz bath, taken once or twice a day at a temperature which feels comfortably hot to the patient, about 100°–110° F, will perhaps cause satisfactory hyperemia Such a bath should not be taken for more than ten minutes and the individual should see that the body and legs are not afterwards cooled It will be found perhaps that the sitz bath works best preceding a period of exercise or that it should follow one before the patient goes to bed

Diathermy will be used by those familiar with it, but since it calls for a special apparatus and involves an expense over and above that of the simple and useful measures already described, it must be regarded as a part of hospital (or office) rather than ambulatory, or home, treatment

Drugs, except so far as they may strengthen the action of the heart, improve appetite, assist sleep or relieve pain, are of no real advantage in the treatment of arteriosclerotic arterial deficiencies

Treatment of Pain—If the patient suffers from spontaneous pain or if necessary exercise causes a serious intermittent limp, special treatment over and above the routine out

lined will be required. But it will first be necessary to discover (1) what actually will relieve pain and (2) whether an effective hyperemia in the limb can be induced. These two considerations can usually be counted as one.

The relief of pain by drugs can be dismissed as something not going to the heart of the matter. Morphine especially is very dangerous (and useless).

A case in point is the following: M.G.L., a man of sixty, had been unable, for many years, to walk at a normal pace without bringing on a cramp-like pain in the front and outer side of the right leg. For some two to three years he had noticed a very similar crampy pain at night. The nocturnal pain had recently increased in frequency and severity. For the last year he had suffered an oppressive (anginal) pain in the midthorax on exercise. This had been benefited by vasodilating drugs.

The patient looked more than his age. He was reasonably well nourished. His legs were only very slightly atrophied. No arterial pulsations could be made out below the knees. There was a good pulsation in the left popliteal artery, none in the right. The right femoral pulse was less strong than the left. Both feet were cool.

Upon elevating both legs for two minutes, the characteristic discomfort in the right calf set in. Both feet also became very pale and on subsequently letting them hang down, the color came back more slowly in the right than the left, not reaching the toes for perhaps half a minute. The X ray revealed faint arteriosclerotic changes in both femorals and definite calcification not only in the dorsalis pedis arteries, but also in the small vessels running to the great toes. While under observation, the patient was given two ounces of whiskey for his pain one night, apparently without relief, yet he did not walk the floor as usual and soon went to sleep.

In a rather warm room,* that is, at a temperature of 78° F. (25° C.) the right great toe was found to have a temperature

* Naturally the test should have been made in a cool room. Unfortunately none was available. Yet the test was reasonably informative.

of 87° F, the left 88° F and in response to a right lumbar block by the injection of procaine, the temperature of the right great toe rose *only 2° F, never catching up with the left,* which also rose a trifle However, the right foot acquired a sensation *of warmth, as compared* with the left, and elevation for five minutes, while the block was effective, failed to bring on the characteristic pain Thus the pain was for the moment benefited, yet with only negligible vasodilatation, a state of things which might have been surmised from the simpler observations already made

In such a case as this, over and above the usual Buerger Allen exercises and the routine protection from cold and trauma, a prolonged trial of suction and pressure or, perhaps better, intermittent venous compression *should be made* So little is to be expected from a lumbar sympathetic resection *as to make this procedure inadvisable,* particularly in view of the cardiac symptoms There is little danger of gangrene, the problem *being to control pain* Drugs will be ineffectual

In the above account it will have been noticed that a right lumbar sympathetic block gave comfort, bringing up the question whether, if routine measures had failed, injection of alcohol into the region of the right upper lumbar ganglia might not properly have been tried Both Flothow and Reichert recommend such a course and it may therefore be proper at this point to describe the procedure

The patient being placed horizontal, lying on the side opposite *to the one into which the injection is to be given,* a wheal is made six to seven cm lateral to the second* lumbar interspace, as in the diagnostic block by procaine (page 27) and after the usual infiltration with procaine of the intermediate sensitive structures, the eleven cm needle is directed, at an angle of about 45°, toward the body of the second lumbar vertebra A second needle is passed toward the third lumbar

* The usual directions are to inject alcohol beside the first second and third lumbar vertebrae but injection of *only L 2 and 3* should secure the *desired* result *and there is* a theoretical objection to destruction of the first lumbar ganglion in the male

vertebra, the plan being to see that the points of both needles are well anterior, that is, toward the front of the body where the sympathetic gangliated chain is situated. After the usual suction, to make sure that no blood vessel is entered and that no cerebrospinal fluid flows, a few drops of ninety-five per cent alcohol had better be injected into each needle. If the points are well placed, a transient epigastric or abdominal pain will be felt. In the absence of this sensation it is almost certain that the needle points are not near the ganglionic chain and they should accordingly be shifted to a more satisfactory position. Once the pain appears, two to four cm. of ninety-five per cent alcohol are slowly injected. In any case, the fluid should be kept well in front of the lumbar spinal nerves, lest an alcoholic neuritis, which may last for a month or two (almost as serious a matter as the original complaint) be set up. Indeed this is the real objection to the procedure which, in many cases, gives a high degree of relief. The first effect of an accurate injection about the ganglia is a severe pain of a few minutes' duration, due to a preliminary irritation of the ganglia and associated with sweating and coldness in the extremity. Following this, a feeling of warmth, both subjective and objective, comes on.

It has been said that smoking in arterial deficiencies should be forbidden. There is perhaps no other routine measure so likely to relieve pain. In the following case, there was only a minor complaint of spontaneous pain, but the intermittent cramp and such spontaneous discomfort as was present were done away with in an almost miraculous way:

S.D.F., a man only fifty years of age, a heavy cigarette smoker and inhaler, had noticed for two years, on walking even for a short distance, a feeling as if a knife had been thrust into the calf of his left leg. This disappeared in the usual way on rest. The trouble had since advanced so rapidly that recently he had been able to play continuously no more than three holes of golf. At the same time he had begun to wake in the morning noticing numbness in the instep and the great toe of his left foot. *The latter* became cold very easily.

The patient looked rather more than his years His blood pressure was rather low (118 systolic) There was an obvious atrophy of the calf muscles No arterial pulsations could be made out below the femorals Both the feet appeared cadaveric on elevation for two minutes and flushed slowly on depression, the left toes requiring twenty seconds before showing color On passing one's hand down the leg, a faint but unmistakable change from warmth to coolness could be felt a little above the ankle The feet, however, were dry and not noticeably cold

The patient was given postural exercises and directed to stop smoking In two months a decided change had occurred. He could now walk slowly on soft ground for two miles without having to stop because of cramp He no longer noticed numbness of his left foot on awaking A faint pulsation could be detected in both the left and right dorsalis pedis arteries, the left the stronger The flush on hanging down the leg after two minutes' elevation passed out upon the toes of the left foot in seven seconds instead of twenty In three months more the patient's cramp had so far diminished that he could walk fast for perhaps a quarter of a mile Then the limp returned He could play eighteen holes of golf

Here is an evident arteriosclerosis with a strong suggestion of superimposed vasoconstriction due to the abuse of tobacco Giving up cigarettes vastly relieved the intermittent limp (postural exercises probably helped) and the peripheral pulses returned, but once this change had occurred, the smaller vessels were found to have little capacity for reactive hyperemia Spinal anesthesia caused the temperature of the great toes to rise moderately, the left 9° F and the right 4° F but in neither case was the low normal level of 90° F (31 5° C) reached

The Treatment of Gangrene —Since pain in arteriosclerotic states is not usually of great importance, the first consideration is the limitation of gangrene, the second, is the securing of healing once the gangrenous part is cast off or removed, and the last, of course, is the problem of amputation In most

instances, gangrene is of the dry or mummifying type, and ulceration is limited and uncomplicated save perhaps by a local necrosis of a phalanx or disintegration of a joint. In the absence of diabetes, infection is seldom serious.

Limitation of gangrene calls for rest in bed. The foot should not be elevated, since arterial blood will then reach the toes with even greater difficulty than is already the case. On the other hand, if the leg is too dependent, edema will occur. It will usually be true that a horizontal position for the legs and a reclining one for the body will give about the right amount of moderate venous congestion and of increased capillary pressure. The foot should be protected from trauma by a large cradle which crosses the entire bed and includes at least the full length of the legs. The cradle should not, however, be heated or if heated the temperature should merely be warm, that is, not over 80°–90°F. For local heat calls for a more active metabolism than the crippled arterial system is able to support and if it does not actually burn the exposed parts, it does them harm (increased anoxemia) rather than good. The leg had better be kept warm in wool or cotton.

The actual dressing of the foot should be made an aseptic ritual [illegible]

usir [illegible]

wip[illegible] with sterile water, or seventy per cent alcohol. Finally, when the gangrenous area has been treated, the nearby skin should be covered with vaseline gauze. For the gangrenous part itself every one is apt to have his favorite remedy. Once securely mummified, its covering, except it be protective and clean, is unimportant. In the earlier stages it may be patted with any mild antiseptic and covered with gauze moist with the same solution. Alcohol, seventy per cent, is useful for its drying quality. The truth is that routine cleanliness is more important than any drug. But if infection makes it seem advisable to keep the dressing moist, one must choose some solution. Samuels warmly recommends soaks of 0.5 per cent watery chloramine solution to aid in the separation of small sloughs and to clean ulcerated areas. A *watery*

iodine solution (Lugol's solution 1–400) or a coconut oil derivative (alkyl dimithyl benzil-ammonium chloride) in a strength of 1–1000 are nonirritating and almost as good dilute antiseptics as any "Eusol",* mixed, equal parts, with mineral oil, is an excellent antiseptic and surprisingly little irritating to the skin

Local Amputation—Once the gangrene is localized and a zone of reaction established proximal to the line of demarcation, or once it is evident that a sinus leads down to a disorganized joint or an area of necrotic bone, the problem of getting rid of the necrotic part and securing healing arises The safest and most time consuming method is to allow a gangrenous toe to be cast off For the tip of the toe this is all very well, but for half a great toe, for a whole smaller one, or for a necrotic phalanx, local amputation, provided certain rules are observed, is economically desirable and may occasionally be performed However, it is almost criminal to amputate such a toe in supposedly sound tissues proximal to the area of reaction Amputation, to be even reasonably safe, must be made very close to the border of the gangrene in the red zone of reaction If the tissues are cleanly divided and not traumatized and if any exposed phalanx is cut across, not disarticulated, the wound can be left open with good prospects of healing Nature has already set up a defense against infection The operation asks little new of the patient

It is at the stage when gangrene is over and granulation plus epithelization are starting that the details of treatment will decidedly help or hinder In well equipped institutions, the Carrel Dakin technique, skillfully used, is effective But treatment of an open wound lined with a thin slough in avascular tissues may include anything from the use of Dakin's fluid to pure urea crystals When a wound is left boat-shaped, as after the necessary removal of a necrotic metatarsal head,

* The formula for Eusol is	Boracic Acid	12 5 grams
	Chlorinated Lime	12 5 grams
	Distilled water	1000 cc m

Mix allow to stand over night and filter

the use of Dakin's fluid or dichloramine-T (eight per cent in chlorcozane) or Eusol and mineral oil are especially recommended. Self-draining wounds are easier to treat.

Once healing is under way, an attempt to secure a reactive hyperemia and a permanent widening of the vascular bed are apt to bear fruit. Rhythmic suction and pressure, or venous compression, will often hasten healing and of course the routine treatment as for the pregangrenous stage of arteriosclerotic deficiency should by all means be resumed. Whether or not any attempt at a permanent block of the sympathetic supply to the limb shall be made is a matter to be decided on the ground of the patient's proven capacity for vasodilatation and the probable estimated future of his circulatory deficiency. On the whole, little is to be expected in arteriosclerotic gangrene from sympathetic resections.

Amputation of the Limb.—It will be profitable to discuss the criteria for the amputations at any level above the toes in connection with diabetic gangrene at the end of this chapter. Amputation is demanded when so much of the foot is destroyed as to make it useless, when a life is to be saved in the presence of uncontrollable infection and, very rarely, on account of pain or for economic reasons. Amputations below the knee can almost never be expected to offer a healthy stump. Most are performed through the knee joint or through the lower third of the thigh.

Thrombosis in Arteriosclerotic Deficiency

Doubtless large vessels already considerably narrowed at some one point can finally be closed by thrombosis without bringing on any sudden or noteworthy change in the circulation of a leg. The rapid closure of an artery carrying a good blood supply is a very different affair. Except that no source of embolism is apparent—that is, the heart is not fibrillating and nothing resembling a coronary infarction has occurred—the suddenness of the ischemia much resembles that of embolism. There will have been perhaps no particular warning, no premonitory numbness, coldness, or limp. The pain of a

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sudden thrombosis is usually severe, sometimes agonizing, but as already explained may take the form merely of numbness or coldness In any case, it is of such a character that the patient is immediately driven to seek help The following is a case in point of a mild sort

C L C, a woman, seventy seven years of age, having previously considered herself well, was seized rather suddenly, three days before coming under observation, with a severe pain in the left leg, radiating downward into the left calf and foot Since then, a burning sensation had settled upon the dorsum of the foot This pain was particularly bad at night, keeping the patient awake as long as the leg was left horizontal in bed but being relieved when the leg was hung out of bed, or when she hobbled about Twenty four hours before entering the hospital, the burning pain spread to the heel There had been no cramps

Examination showed a rather frail old woman, evidently suffering On exposing the legs in a cool room, the right foot retained its heat fairly well The left foot cooled rapidly and noticeably, as compared with the right, up to a point some three inches above the ankle joint When the legs were allowed to hang straight down, the toes of the left foot soon turned reddish blue (the right also to a lesser degree) and this color gradually faded out upward toward the ankles No pulsations could be felt over either dorsalis pedis artery There was a questionable pulse in the right posterior tibial—practically speaking no pulsations in either leg below the femorals at the groin The X ray revealed calcification in both posterior tibial and dorsalis pedis arteries

The greatest amount of comfort was secured by allowing the old lady to sit in a chair, the legs resting on a stool at a slightly lower level In this position and with only the aid of mild sedatives, she passed fairly good nights, whereas if she attempted to spend the night in bed, she was made sleepless by pain, which was always of the same burning sort and over which drugs, even opiates, had so little control as to be almost totally useless

Suction and pressure gave considerable additional relief. It was applied, at first rather tentatively, for half an hour twice a day, and with such improvement that it should undoubtedly have been used more freely, but like most elderly women, this one was utterly intolerant of hospital care. She refused any further treatment which necessitated her stay in the hospital.

Upon going home she deteriorated rapidly: her pain increased, she lost her appetite, and lived only for about a year. She never developed actual gangrene.

Some thromboses cause such sudden widespread ischemia as to be fatal, the effect of the closure being shocking and attended by other signs of circulatory failure and death within a few days. In such cases the foot and half the lower leg often become gangrenous.

Less serious accidents, yet leading to gangrene, may resemble the following:

P.N., a woman, sixty-four years of age, had been well up to three weeks before coming under observation. Then, rather suddenly, the right leg began to "feel sore" and she found that she must lie down after walking only a very short distance. The right foot almost at once became blue, the toes black. Coldness and numbness "bothered her greatly". Vesicles soon appeared upon the lower third of the leg and foot, increasing in size as the discoloration spread upward.

Examination showed a well-preserved woman. No pulses could be felt in the right leg below the femoral, whereas the pulsations in the left leg, including both posterior tibial and dorsalis pedis, were normal.

The right leg, as can be seen in Plate I C was the seat of what seemed to be a moist gangrene. Yet the patient was treated conservatively, sterile dressings being applied, and on leaving the hospital about a week later, against advice, the lower leg and foot were shrinking and the cyanotic, cold area was receding. Thus the threat of infection, especially with the gas-producing bacilli, was being removed. The patient has since been lost sight of, yet it hardly seems possible that she

saved her foot In such a case as this, careful study of the level at which an efficient circulation ended, might have permitted an amputation below the knee

The fact that gangrene starts in a moist way does not necessarily call for an immediate amputation Yet the danger of serious infection is such that the leg must be watched intensively If infection shows itself and spreads upward, an emergency guillotine amputation will be required With a neat, clean amputation, at or even below the knee, as the reward if conservatism succeeds, and an emergency guillotine amputation, possibly at a high level, the penalty of failure, the difficulty of treating a case of this sort is apparent

Diabetic Gangrene

Diabetic gangrene is really arteriosclerotic deficiency plus actual or potential infection in a diabetic The background of arteriosclerosis is only partly the effect of the diabetes (lipemia, deposit of fat in the media of the arteries) for arteriosclerotic deficiencies in diabetics show themselves only a few years earlier than do similar deficiencies in nondiabetics It is really the lowered resistance to infection, especially by the common pyogenic bacteria of the skin, which gives diabetic lesions of the toes and feet their especially dangerous character In 1925, Coller and Marsh called attention to the importance of separating the pure arterial deficiencies from those primarily due to infection This McKittrick has followed up, presenting the clinical picture under the heading of "Conditions due primarily to arterial insufficiency" and "Conditions due primarily to infection" This classification will be used here but it should be kept in mind that any but really young diabetics are likely to be arteriosclerotic, and that even in those whose disorders seem typically arteriosclerotic, the tissue cannot be expected to have even the ordinary arteriosclerotic's resistance to infection

Conditions Primarily Arteriosclerotic —*The onset of symptoms is not different from that of a nondiabetic, arteriosclerotic deficiency except that the patient may be five years*

younger. Coldness, numbness, intermittent limp are common complaints. The symptoms calling for treatment will seldom have been present for more than a few months, and usually for weeks rather than months. The appearance of the toes and feet is like that of arteriosclerosis. There will often be left some portion of the peripheral arterial pulsations. There is the same liability to closure of a large artery by thrombosis, and there is even more of a tendency to irritation by a deformed nail, to infection of a badly treated corn or callus. Thus the routine treatment, outside of that required by the diabetes itself, is the same—encouragement of the circulation in general, preservation of the warmth of the limb, vascular exercises, and in particular, exquisite care of the nails and skin.

When gangrene or ulceration or necrosis of some of the toes occurs, it looks and behaves like any arteriosclerotic lesion. All diabetics with a poor peripheral circulation are not sitting on powder barrels! Some have a mild disease controllable by diet, and even a condition bad enough to require insulin does not necessarily rob the individual of *all* resistance to infection. The real trouble is that when infection once starts, fixing itself upon a joint, entering a tendon sheath, and in particular progressing along the lymphatics up the foot and leg, there is set up at once a vicious circle of deficient circulation, lowered resistance to infection and aggravation of the diabetic state which will often take life if it is not broken by a radical amputation. Undoubtedly, a fatal infection can, though it seldom will, start from a perfectly dry gangrene of the outer half of the toe. However, as an example of what [illegible]
in how thi [illegible]

J.Z.Q., [illegible] of sixty-seven years, had [illegible] well
[illegible]
ti [illegible] this
o [illegible] was
[illegible] to take vasodilating drugs. At the same period, a diagnosis of diabetes was made and at one time insulin was required. Yet in the succeeding years the disease

made so little progress that he was not even very careful to follow his diet When he entered the hospital his blood sugar was averaging 0 215 per cent, his urine showed only a trace of sugar, and he soon became sugar-free upon diet alone, the blood sugar remaining at 0 146 per cent

Ten days before entry, the left great toe was noticed to be dark in color, since when a sore, inflamed area had gradually developed upon the inner edge of the great toe nail The toe had become increasingly painful and tender

The patient was a fairly well preserved, cooperative individual whose peripheral arteries were everywhere hard. There was a suspicion of an arteriosclerotic aortitis All pulsations in the right leg, including both the dorsalis pedis and the posterior tibial, were present, those on the left below the femoral were absent (recent thrombosis?) The left great toe was deep red An area of excoriation was present along the inner edge of the left great toe nail, from which a little pus oozed Under alcohol applications and hot saline dressings, the cellulitis promptly cleared up The nail was trimmed square but not short A small area of granulation was left, but the whole toe from the metatarso phalangeal joint down remained a deep red without actual gangrene

At this point a mistake was made Instead of insinuating a little gutta percha tissue or some other nonirritating substance under the edge of the nail and continuing the warm wet dressings, the nail was avulsed under *spinal anesthesia* This left still more exposed raw surface, which, during the next few weeks showed no tendency to heal Here surgical action was again substituted for conservative treatment

Instead of continuing antiseptic dressings and instituting attempts at reactive hyperemia, which might have encouraged granulation and epithelization, the toe was amputated, not in the zone of redness, but above it, not by dividing the proximal phalanx, but by disarticulating the toe at its base Result, a sloughing wound, but fortunately no extension of infection A month later there was evidence of circulation in the skin about the sloughing hole, and a little new skin had grown about its

edge. The battle is therefore a draw. Very likely the patient's leg will be saved. With a greater display of patience his toe might have been!

The subject of conditions due primarily to arterial insufficiency can be summed up by quoting, with explanations, from McKittrick and Pratt.

1. No or feeble pulsations are present in the peripheral blood vessels of the leg.

2. The foot is cold, blanches on elevation and becomes dusky or red and shiny when dependent. Between the mid-foot and the knee there is frequently a level at which the cutaneous temperature can be felt to change.

3. When necrosis of some part of the bony structure of a toe occurs, gangrene is usually evident.

4 Pain is apt to be more severe than the local lesion appears to warrant.

5. Gangrene is common

Local amputations in this group are rarely successful and frequently are dangerous. Infection in a pulseless foot is the origin of most cases of septicemia. Thus, in the pregangrenous stage, the usual routine treatment, as for any arteriosclerotic deficiency, should be used, special care being given to avoiding minor infections about the nails. When gangrene is present, the toe or area should rigorously be protected, and in most cases allowed to separate. Such treatment is described in a later section.

Conditions Due Primarily to Infection.—Whether or not gangrene is present, infection in the form of cellulitis and lymphangitis, that is, a streptococcal form, is obvious. A favorite initial lesion is the ingrowing toe-nail, runaround or ill-treated corn or callus. One toe, or a toe and part of the forefoot, is red and swollen. One or more red streaks of lymphangitis are apt to extend over the foot, upon the ankle and even up the lower leg. Pulsations in the vessels of the foot are often vigorous. Pain is not a feature and the foot is warm, even hot, partly because the arterial circulation is sufficient but more because of the inflammation. To sum up:

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duration about the base of the toe. The flexor tendon was sloughing, the sheath infected and discharging. Though pulsation was present in the dorsalis pedis artery, the X ray showed the local small vessels to be calcified. Under alcohol applications and hot packs, a line of demarcation became clear and there developed a zone of reactive redness proximal to it. The great toe and the partly necrotic head of the first metacarpal bone were then successfully removed under spinal anesthesia, the amputation being carried out close to the gangrene through the red swollen tissues, that is, in the zone of reaction. Result, good healing. However, within two years gangrene of both feet followed, probably because the diabetes was neglected.

Treatment of Diabetic Gangrene: *Arteriosclerotic Type.*—It has already been stated that gangrene due to arterial deficiency should be treated conservatively, partly to avoid dangerous infection, and partly because local amputations will seldom result in healing, even if no infection follows. Precautions against infection being taken, the gangrenous part will often be cast off. Then, under antiseptic dressings and vascular exercises, healing is likely to occur. If economic reasons and the likelihood that the foot, even if healed, will be disabled urge an early amputation of the leg, the operation should be performed at the knee or in the lower thigh, depending on whether or not the individual is or is not likely to be able to use an artificial leg.

Infected Type: Emergency Treatment.—Provided the nature of the local infection—cellulitis, osteomyelitis of a phalanx, suppurating joint or tendon sheath—offers a reasonable hope that local treatment in the form of hot wet dressings, local drainage or local amputation will be curative, blood should at once be drawn for a study of the blood sugar and to see if bacteriemia can be demonstrated. Cultures should also be taken from the local lesion. Medical and surgical treatment must from the start go hand in hand. The use of insulin should at once be begun. Conservative treatment should be carried out tentatively for a preliminary twelve-hour period.

If, after twelve hours, local conditions are no worse and the

1 Pulsation in the dorsalis pedis artery is usually present

2 The *foot* is warm and of good color Even if the heat of the inflammation is not present, the observer's touch will find no marked change from warmth to coolness in passing the hand downward from knee to foot

3 Necrosis of the bony structure of a toe without gangrene is very common

4 Pain is related only to the infection and is usually less than the *local condition* would lead one to suspect The part may even be remarkably anesthetic

5 *Gangrene* is found only in the presence *of infection or after* trauma

The following are illustrative cases

H S C, a male fifty-nine years of age Story of diabetes for one year, to which little attention had been paid For two weeks, a *black slough had been present* over the right great toe joint Beneath this was fluctuation and a disorganized metatarso phalangeal joint There was redness and swelling of the surface of the foot back to the instep *Good pulsation* in the dorsalis pedis artery No pain Insulin, fifteen to twenty units a day, barely controlled the diabetes Incision, under gas oxygen anesthesia, showed infection of the deep fascial spaces of the foot and failed to halt the infection Amputation (closed) through the leg, six inches below the tibial tuberosity, was followed by healing but the stump was never satisfactory A better and safer procedure would have been a guillotine am *putation through the mid-leg followed* by a closed amputation through the knee joint (Gritti Stokes or Callander type) Very likely an initial closed amputation through the knee would have succeeded *Such operations* give *a very good stump* and the chances are that this patient should be able to use an artificial limb with success

E C, a woman, forty seven years of age, a diabetic for two years taking fifteen units of insulin a day For seven weeks she had suffered from an infected blister on the outer surface of the right great toe During this time the great toe had gradually turned black and finally mummified The blood sugar on entry was 0 178 per cent There was redness and in-

toes (all of which end about opposite the metatarsal heads). These should be entered from a lateral incision as shown in the accompanying sketch.

For cavities having no dependent drainage, the use of Dakin's fluid, with all the Carrel-Dakin ritual, is very satisfactory. Indeed this elaborate technique finds here its greatest usefulness, and the various allied solutions and oily preparations are only slightly less valuable. The skin of the foot,

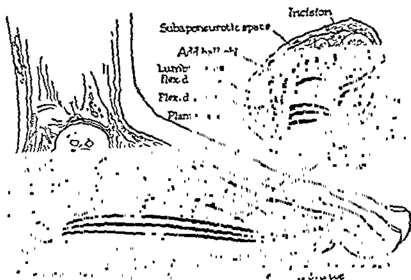


FIGURE 5. THE FASCIAL SPACES OF THE FOOT—(modified from Grodinsky. By courtesy of *Surgery, Gynecology and Obstetrics*, 49·737, (Dec.) 1929. From Homans's *Textbook of Surgery*. Courtesy of Charles C. Thomas, Springfield, Ill., and Baltimore, Md.).

which is so easily injured, must be carefully protected with vaseline gauze. No dressings can be allowed to adhere to raw surfaces. If no antiseptics are used on self-draining wounds, the edges of the skin should be protected with a smooth substance such as gutta percha tissue or vaseline gauze.

AMPUTATION

Since no artificial knee joint can compare with one's own, the ideal amputation is one which leaves a four to six inch stump below the knee. Such a stump, from the very nature of

patient's diabetes seems controllable, a further twelve hour period of observation is permissible At this time the evidence of the blood culture and the nature of the local infection will be available The use of sulfanilamide (or an equivalent drug) is very likely to be indicated by the bacteria present In that case, administration of a full dose of the drug, to secure the optimum percentage in the blood, is required

If the blood culture is positive, the local lesion is no better or actually worse and the blood sugar difficult to control, a major amputation should be performed

If, on the other hand, all conditions seem favorable, the local treatment is continued with the idea of performing a minor amputation or operation for drainage at the appropriate moment

Under such a system, and provided it is thoughtfully planned, local treatment by amputation or drainage will usually succeed That is, if one third, for instance, of all infected diabetic gangrenes are treated conservatively, only a small proportion of these should require a major amputation to save life or because of failure to secure healing The other two thirds—the figures are intentionally vague since conditions and skill are variable—will require a major amputation on sight, or at the end of twelve to forty eight hours of expectant treatment, or following failure of a local amputation to halt infection

Local operations to control sepsis, whether or not they include the amputation of toes should not be performed in the presence of an ascending lymphangitis, but should await its subsidence

They should take advantage of every bit of local defensive reaction which warm moist antiseptic dressings, immobilization, and slight elevation are able to produce If a toe is to be amputated, incisions should be kept away from the sole of the foot If a metatarso phalangeal joint is infected, the metatarsal head will usually be disorganized and its removal will make drainage more effective The fascial spaces of the sole which Grodinsky has described are very often invaded by progress of infection from the flexor tendon sheaths of the

The Temperature of the Skin.—The methods of determining the skin temperature and the inferences to be drawn from such observations are described in Chapter I. In applying them to the problem of selecting a level of amputation, gross evidence is secured first by exposing the previously warmed leg to a cool atmosphere. If one leg from the knee down is cool to the touch, as compared with the thigh and the other leg, any amputation below the knee will be out of the question and even one through the knee joint will be dangerous. Such an observation can of course be confirmed by the thermocouple and by oscillometry. A very satisfactory observation is the McClure-Aldrich test, which consists in making cutaneous wheals with 0.2 ccm. of physiologic saline solution. Normally these wheals should remain visible for the better part of an hour. They are made in series downward from the upper thigh, at four inch intervals, as close to the foot as seems worth while. In the parts poorly supplied with blood the wheal may last only a few minutes. Thus the lowest wheal which lasts more than the half hour marks fairly well the low limit of any proposed skin flap and the bone will of course be divided at a considerably higher level. The wheal can also be made with 0.1 ccm. of a 1–1000 solution of histamine (in one per cent *novocaine*), the normal reaction being a hyperemic flare and a wheal, but this test probably possesses no particular advantage.

Palpation of the Arteries.—A dorsalis pedis or posterior tibial pulsation suggests that an amputation below the knee will probably be successful. However, it does not guarantee the result. Sepsis, for instance, may forbid the operation, or the patient's history of intermittent limp and numbness of the foot may prove that it is the finer circulation, on which the nutrition of the flaps depends, which is lacking. In other words, arterial pulsation alone is not enough. Other tests must be in harmony.

A popliteal pulsation guarantees nothing for the foot. The main vessels below this point may be defective. Again, not only is the normal pulsation difficult *to feel* in many persons,

the disease—arteriosclerotic deficiency—for which most of the amputations under discussion here are performed, is almost impossible to secure Only upon the evidence presently to be described should the mid leg amputation be considered.

The next available level is that of the knee joint Here amputat

tion,

the anastomotic vessels about the joint The femur is usually divided through the upper part of its flaring condyles and the prepatellar tendon (the patella being excised) or the refreshed posterior surface of the patella itself is used to cover the cut end of the bone, giving what often proves to be an end bearing stump Amputations of this sort are invariably closed and are intended to be used for an artificial leg having an artificial knee joint This joint, and indeed the use of the artificial limb in general, requires, on the patient's part, normal sight, normal balance, and normal strength, in fact a moderate athletic ability And two such legs call for a very able bodied, courageous, youthful person

The amputation through the lower third of the thigh is a procedure primarily safe The stump which it leaves will support an artificial limb but very often it is performed with little idea that a limb can be worn

The guillotine amputation—an emergency procedure to save life threatened by infection—is almost always made in the leg, through its mid-portion, a circular division intended to be left wide open, its surface flat Such an amputation can rarely be trimmed by a plastic procedure to leave a permanent stump below the knee Almost invariably it is followed by an amputation through the knee or lower thigh Rarely a guillotine amputation is made in the lower thigh, with the object, first, of saving life, and second, of leaving a stump which will be closed by a plastic but will probably not be required to support an artificial leg

The Selection of a Level for Amputation is made with the above considerations in mind The following tests should be used

The Temperature of the Skin.—The methods of determining the skin temperature and the inferences to be drawn from such observations are described in Chapter I. In applying them to the problem of selecting a level of amputation, gross evidence is secured first by exposing the previously warmed leg to a cool atmosphere. If one leg from the knee down is cool to the touch, as compared with the thigh and the other leg, any amputation below the knee will be out of the question and even one through the knee joint will be dangerous. Such an observation can of course be confirmed by the thermocouple and by oscillometry. A very satisfactory observation is the McClure-Aldrich test, which consists in making cutaneous wheals with 0.2 ccm. of physiologic saline solution. Normally these wheals should remain visible for the better part of an hour. They are made in series downward from the upper thigh, at four inch intervals, as close to the foot as seems worth while. In the parts poorly supplied with blood the wheal may last only a few minutes. Thus the lowest wheal which lasts more than the half hour marks fairly well the low limit of any proposed skin flap and the bone will of course be divided at a considerably higher level. The wheal can also be made with 0.1 ccm. of a 1–1000 solution of histamine (in one per cent novocaine), the normal reaction being a hyperemic flare and a wheal, but this test probably possesses no particular advantage.

Palpation of the Arteries.—A dorsalis pedis or posterior tibial pulsation suggests that an amputation below the knee will probably be successful. However, it does not guarantee the result. Sepsis, for instance, may forbid the operation, or the patient's history of intermittent limp and numbness of the foot may prove that it is the finer circulation, on which the nutrition of the flaps depends, which is lacking. In other words, arterial pulsation alone is not enough. Other tests must be in harmony.

A popliteal pulsation guarantees nothing for the foot. The main vessels below this point may be defective. Again, not only is the normal pulsation difficult to feel in many persons,

but the arterial circulation below a slowly closed femoral artery is sometimes remarkably efficient A more significant finding would be the sudden disappearance of the popliteal pulse, in which case the femoral artery must rapidly have become plugged With such a change, the clinical signs would undoubtedly agree, as in a case quoted earlier in the chapter, and any other than a guillotine amputation could never be performed below the knee A feeble femoral pulsation at the groin forbids amputation below the lower third of the thigh

Arteriography has been used in some clinics to indicate the level at which an amputation may be expected not only to heal cleanly but to leave a well nourished stump Until the skill required for its authoritative interpretation is more generally distributed, simpler methods should be trusted

Amputation—The various operations cannot be described here It is enough to discuss the present day tendencies Most amputations below the knee are emergency guillotine procedures, that is, circular divisions performed below an Esmarch bandage The tibia should be divided four to six inches (ten to fifteen cm) below its tubercle, flush with the retracted muscle In the ordinary closed amputation, the fibula would be divided one to two cm higher but for the guillotine the matter is not important

For the finished procedure it is a matter of indifference in these days whether the scar is at the tip of the stump or the side The main thing is to draw some muscular or tendinous structure over the end of the tibia, and the skin flap may well come mainly from the same direction Thus, the musculocutaneous flap comes best from the lateral and posterior faces of the calf Bulky muscle should not be used, but its aponeurosis, and perhaps some muscular backing, makes a useful covering for the divided end of the bone

The amputation at the knee joint, according to the tendencies today, is made at the point where the condyles begin to flare The skin flaps are rather long For the Gritti Stokes, the anterior flap includes the skin in front of the tibial tubercle, the posterior flap being shorter The patella and its tendon are

saved, the posterior face of the former is sawed off, and it is fastened to the open end of the femur. Here it is expected to heal, but does not always do so. If it heals, an excellent end-bearing stump results. A rather simpler and more generally acceptable procedure is an amputation at the same level, by which the patella is removed and its tendon made fast over the end of the femur. This gives about as good a stump. The lining of the knee joint is not removed. Callander's operation is of this sort, but it is peculiar in several respects. That is, the prepatellar tendon is merely laid over the end of the bone, the posterior flap is left very long, and nothing is sewed together, the skin flaps being kept from separating only by a few clips. Limbs having a very unpromising circulation can be amputated in this way. The posterior flap draws back, the tendon heals over the bone, and a good stump results.

The thigh amputation exposes large muscular surfaces, which in themselves have little power to heal, but to balance this the skin is now cut in a nearly circular way, so that the operation is not only well away from the infected or potentially infected field, but the superficial tissues are given the ideal opportunity to heal. For no flap can be as well nourished as the skin adjacent to a circular cut.

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CHAPTER III

THROMBO-ANGIITIS OBLITERANS

THIS disease, since its first recognition, has been noteworthy as having a peculiar etiology. It is decidedly more common among Hebrews, especially of Polish or Russian origin, than in any other race. It is nearly confined to males. And it is so greatly aggravated by the smoking of tobacco that really serious gangrene is hardly seen in others than heavy smokers. It is not purely a disease of arteries. This von Winiwater recognized in his excellent description of a leg amputated by Billroth for gangrene in 1877. He described a "Peculiar form of Endarteriitis and Endovenitis with Gangrene of the Foot", thereby calling attention to the involvement of the veins as well as the arteries and separating the disorder from arteriosclerosis. Since his day, superficial migrating phlebitis, which comes and goes so often with the fluctuations of the disease, has been added to the picture; and finally, in 1908, Buerger, identifying the characteristic pathology with the symptom-complex of spontaneous presenile gangrene, gave the name "Thrombo-angiitis Obliterans".

Buerger holds, "that the disease begins with an inflammatory lesion attended with occlusive thrombosis, and that it affects the arteries and veins in a sort of relapsing fashion, very much in the same manner as in the veins in migrating phlebitis". In other words, it takes the form of repeated attacks, during which new groups of vessels are affected, alternating with remissions, during which organization occurs and a collateral circulation is established. With others, he believes that the acute reaction which the superficial veins exhibit in a minor percentage of all cases, offers the only practical means of studying the earliest stage of the disease, for by the time

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thrombosis were primarily peripheral and tended to extend centrally, a very interesting point in view of present-day experience. For many observers feel that the most malignant form of the disease is one which shows itself primarily in the peripheral, or smaller, vessels, as contrasted with a more tractable form which tends to an early occlusion of the femoral itself. It seems to be the peripheral form which causes most intractable pain, is less easily circumvented by a collateral circulation, and is most apt to lead to amputation. Whereas the more central occlusion is not only less painful but is rather rapidly relieved, for the time being at least, by a collateral circulation. Now Buerger's studies were made on the amputated legs of the most malignant type of disease, so that he may not have been describing the commonest form. However this may be, Buerger established the tendency of the disease to attack and gradually close the anterior and posterior tibial arteries and veins, causing them to become adherent to each other and in many cases to engulf in the inflammatory process the associated nerves. And since not only peripheral nerves but sympathetic fibers as well must often be affected, it is easy to understand how the pain of gangrene in thrombo-angiitis obliterans is so often far more severe than that of the arteriosclerotic type and why some degree of vasomotor spasm should so frequently be present.

The thrombosis of the early stage of the disease probably results in a greatly diminished caliber rather than permanent obstruction of the arteries and veins. Organization is followed by canalization and the circulation is resumed through narrowed vessels. From a clinical point of view, it is only the arterial disease which is of consequence. Doubtless the extent of the inflammation and thrombosis varies widely according to the severity of each attack in different individuals. But as the years go on, the arterial tree, in the whole limb, is gradually narrowed, the place of its larger vessels being taken by countless fine collateral twigs. Thus it is rare that the peripheral pulses should survive for many years, the upper femoral alone being detectable. This state of things was long ago rec-

the deep vessels can be examined (after amputation) only the late changes can be seen However, no examination has ever revealed the nature of the etiological factor, though typhus fever, ergot poisoning (from rye bread), and a hypothetical recurring vascular spasm have from time to time been looked to hopefully as causes

Buerger describes an acute inflammatory process involving all coats of the vessels, and pictures foci containing "giant cells, endotheloid cells or angioblasts and numerous broken down leucocytes", showing actually a purulent area in a thrombosed internal saphenous vein Yet though various parts of the walls of both veins and arteries have been found in some degree infiltrated with polynuclear leucocytes, most pathologists have been unable to state positively whether thrombosis precedes inflammation or inflammation, thrombosis

The gross lesion is decidedly thrombosis To quote Buerger again "Usually the vessel is seen to be filled with a grayish or yellowish mass that can be distinctly differentiated from the annular wall of the vessel, and that appears to be pierced at one point (more rarely at a number of points) by an extremely fine opening through which a minute drop of blood can be squeezed Such an obturating lesion is firmer in consistency and does not at all resemble the crescentic or semilunar occluding masses typical of arteriosclerosis The vessel is usually contracted so that its wall appears somewhat thickened" Apparently the length of vessel involved varies greatly In the case of an artery, for instance, the process, at its proximal limit, may cease suddenly, the vessel above that point being entirely normal Below, the red clot may show a long conical end So far as the veins are concerned, thrombosis is coextensive with that in the arteries Actually, venous thrombosis is entirely overshadowed by the arterial disease, and so solidly fixed that embolism never occurs

In some of Buerger's first (eleven amputated limbs) specimens the process extends "from the dorsalis hallucis almost into the popliteal" Indeed he speaks as if inflammation and

Migrating Phlebitis is seen in the arms quite as often as in the legs. Indeed it can be studied most easily here. There is an obvious but very mild inflammatory element in every attack. Some vein upon the back of the hand, for instance, becomes slightly painful and sore to the touch. The overlying skin is sometimes reddened. The vein itself, for perhaps two to five cm. (an inch or two) of its course, is noticeably thickened, partly because of hardening of its walls and partly because of adherence of the surrounding soft parts. Yet the skin over the vein is hardly warm. After a few days, such soreness as has been present disappears, and in the course of a week or two the thickened vein itself becomes difficult to feel. At about this time, a new stretch of vein, central to the first but not continuous with it, often becomes sore, or the opposite limb is attacked. Rarely are more than two or three short stretches upon the arm affected in any one period, but it is not so very unusual, in an individual who has become wonted to a migrating phlebitis, to notice the involvement of a vein upon the temple or scalp.

Excision of a vein in the course of a wandering phlebitis *reveals* the typical inflammation of all the vessel's coats. It is the writer's impression, from a rather limited experience, *that* the infla[illegible]s, which indeed [illegible] process finally subsides, leaving behind a vein no longer noticeable and seemingly functioning.

Visceral Manifestations.—It has been asserted and indeed it appears to be true that rarely and only in the most chronic and persistently vicious sort of thrombo-angiitis *obliterans*, thrombosis in some *form may* attack the mesenteric vessels, giving rise to peculiar and annoying attacks of colic. Apparently the involvement of vessels is not extensive enough to cause gangrene, nor is any particular site for the process recognized. Attention may be called to either the large or the small bowel. There is neither fever nor distension. The colic comes and goes, being *excited*, perhaps, at one time by eating and at another by defecation!

ognized from the pathological side, but the clinical picture has always been consistent with it and it can now readily be demonstrated by arteriography

Thrombo angiitis obliterans is rarely acute enough to cause early or extensive gangrene The arterial supply, delivered by countless fine vessels, may be inadequate for an active life, but is not often so deficient as to cause a necrosis of more than one or several toes For the same reason, the peripheral parts in Buerger's disease have, as compared with arteriosclerotic toes, a considerable resistance to bacteria. They can often be lopped off one by one, so to speak, at a very moderate risk of infection Long before a spot of gangrene appears under the edge of or beside the great toe nail, that toe and perhaps others will usually have shown the cyanosis, swelling, and shiny atrophic skin of a slow and much restricted circulation Gangrene of one or more toes and even part of a foot does occur with a fair degree of frequency as the disease advances, but it has been said with a good deal of truth that it is not gangrene which calls for major amputations but uncontrollable pain

Thrombo angiitis of the Upper Extremity—The disease only attacks the arms in the severer cases and after showing itself for many years in the legs Less is known of its pathology in the upper extremities than in the lower, since outspoken gangrene and amputation of more than fingers or finger-tips are exceedingly rare However, obliteration of the larger arteries, particularly the radial, does occur, and apparently the small vessels supplying the fingers are rather irregularly involved The tip of one finger or more will occasionally turn white or cyanotic, showing thickened skin under the nail Sometimes a whole hand will be affected, becoming bluish and cool But there is a decided tendency toward the *opening* up of new pathways and rather more evidence of vasomotor spasm than is likely to be noticed in the toes and feet Even in the severer forms of the disease, amputation, successfully, of several fingers or finger-tips, without loss of the hand is the worst which the patient is likely to experience

tion. Such a change is not necessarily painful, though spontaneous pain may from this time set in before any actual gangrene occurs. Along with the redness or blueness, the skin is apt to be shiny, the forefoot a little swollen, a state of things which the patient's desire to keep on his feet will often exaggerate. Moreover, probably because blood most easily reaches a dependent part, the foot is usually more comfortable when dependent, so that the individual must get up at night and hang the leg out of bed. Thus a vicious circle of deficient circulation and edema is set up, the edema diminishing the already restricted arterial flow and so adding decidedly to the patient's troubles.

By the appearance and behavior of the toes, better than by any other factor, the course of the disease can now be traced. Following the onset of painful reddish blueness and edema, some one toe or part of a toe may become purple and then black, the gangrene being of the dry or mummifying sort. Very often, as the gangrenous part becomes demarcated and is finally amputated, the appearance of the other toes improves. Further attacks sometimes follow, by which other toes are lost, but in the intervals the extremity is of a reasonably good color, the remaining toes limber, free from swelling, and not unduly cold. Such a state is shown in Plate II B. In other and more serious cases the toes remain cyanotic, cold, and shiny. Pain becomes severe. It will perhaps have a burning, agonizing quality and with it will go a high degree of hypersensitiveness. No one must touch or move the patient's toes, but he will sit in bed gently kneading his foot between his two hands. Such a state may last for months, the appearance of the extremity remaining unchanged. Dependency gives some relief but increases edema and so works against improvement in the local blood supply. Finally, if and when the pain is conquered, the toes are left discolored, stiff and useless, some of the nails so deformed perhaps as to cut into the flesh and threaten local infection. Evidently such a patient is hardly better off than if his toes were gangrenous, and indeed his future will actually be made safer if, at a favorable moment, the toes are removed,

Clinical Manifestations.—Buerger backs up his contention that an inflammatory reaction ushers in the disease by calling attention to certain premonitory symptoms before the peripheral arteries give evidence of obstruction. Such are, "lancinating pains in the legs, especially in the calf and foot, cramp-like pains in the leg, first interfering with walking, later requiring complete rest, tenderness in the calf and along the anterior tibial region, simultaneously with, preceded by, or unassociated with attacks of migrating phlebitis." Such observations must be unusual and represent a very acute form of the disease.

Intermittent claudication is usually the initial symptom. Its pathologic basis has already been described (Chapter I). Clinically, the pain complained of may be a sort of numbness, or numbness followed by cramp, or pure cramp or even a feeling as if a knife were being thrust into the leg. And it may be felt in the mid calf or in the anteroexternal group of muscles of the shin or even the foot. Sometimes the pain, at its height, ends by moving into the back of the upper thigh and buttock as if the sciatic nerve were affected. But always about the same amount of exercise brings on the pain or disagreeable sensation—walking so many blocks at such a speed. The more fully the pain has been allowed to develop the longer will be the period of rest required for its relief. Walking on a brick sidewalk brings on the limp more quickly than on the grass. It is more troublesome in cold weather than hot. When the pain is on, the leg is a little sensitive to touch, but not obviously changed. A limp is likely to appear suddenly. Very rarely, in its early stages, it may come and go, in which case a vasomotor element may enter into it, or perhaps a sudden alteration in blood pressure. But as a rule, an intermittent limp, once present, has come to stay.

After months, or it may be years, thickened skin often appears under the toe nails, especially upon the great toe, the nail itself taking on perhaps an unnatural transverse curve. At this time some of the toes, again the great toe in particular, are apt to appear red or reddish blue in the dependent posi-

morning, these toes would turn purple but after walking for a while their normal color would return and the burning discomfort would be for the time relieved. In the next two months, the toes, with the exception of the middle one, improved. This, however, grew more discolored, and "there is a burning feeling and also a pressure feeling as if the toe were being crushed". A trophic disturbance in the form of excessive peeling of the skin from the other toes was evident. The left foot sweated excessively. Hot water upon the skin was intolerable. Pain was so severe at night that sleep could only be secured when the left leg was dependent, so that the patient had come to sleeping in a chair. Occasionally he noticed a cold, stiff feeling in the finger-tips of both hands.

Examination showed a young man of sanguine complexion, evidently suffering. No pulsations could be made out in the peripheral vessels of the left foot. The skin of the toes sweated constantly. It peeled freely from the great and little toes (fungus infection?). The color of all the toes but the middle one was normal enough but all were hyperesthetic. The patient feared to have them touched. The left middle toe was purple, its outer portion black, dry, and shrunken.

Amputation of this partly gangrenous toe through the metatarso-phalangeal joint left a dry reactionless wound which failed to heal. Meanwhile the burning pain grew worse and other toes began to show patches of gangrene, notably the great and second.

A month after the local amputation, the patient's state was pitiable. Pain was continuous. The amputation wound, though very little infected, was unhealed. The outer half of the great and second toes was now gangrenous. On the ground that this was Raynaud's disease, some observations of reactive hyperemia in response to the application of an Esmarch bandage to the thigh were made but were not followed up. The foot flushed slowly, the toes not at all.

Amputation was performed six inches below the tibial tubercle, a closed procedure which entirely relieved pain and which healed ideally. Pathological examination showed that the pos-

a prophylactic sort of amputation occasionally used by those very familiar with the disease

The gangrene of thrombo angiitis obliterans, when it occurs in such a foot as has just been described, starts in a small way, beside a toe-nail, beneath a callus, in the outer half of a toe Indeed, it resembles the arteriosclerotic sort, except that there is apt to be more cyanosis and swelling of the other toes and near by foot Above all there will usually be far more pain and that of the characteristic agonizing sort In the most serious cases, more than one toe, even a part of the foot is apt to become necrotic Pain and hypersensitiveness are sometimes relieved by amputation of the gangrenous toe or toes but more often are not Under these circumstances, the wound of a local amputation often fails to heal, becomes the starting point for more local infection and leads to an extension of gangrene Such is the worst form of the disease, the sort which so often ends in amputation of the limb

The following are cases illustrating respectively a very acute, malignant form of the disease, a serious sort, yet more amenable to treatment, and a variety so tractable that it can almost be considered to have been cured Smoking is an obvious factor in all three The cases were not treated upon any consistent plan They are selected because a good deal of positive information about them has been secured, permitting the clinical signs and pathologic background to be compared

W J D, twenty one years of age, a Massachusetts born Irish American, came under observation in 1916, complaining of a sore toe He was a vigorous young fellow who admitted smoking only fifteen to twenty cigarettes a day The patient's race, the date, and the smoking are emphasized because at that time it was hardly believed that thrombo angiitis obliterans could occur in any but Hebrews or that the cigarette could be so serious an aggravating factor as it is now believed to be The patient continued to smoke throughout his illness

For a vague period, both feet had felt cold, the left more than the right For five months, a burning sensation had been noticed in all the toes of the left foot On first standing in the

About the next case, distinctly less acute, much information was secured by surgical methods which should be condemned for general use, and which were here more successful than they deserved to be.

W.J.S., a Massachusetts-born man, twenty-eight years of age, without Jewish blood but a cigarette and pipe smoker, had suffered for about eight months before coming under observation from "tightness and pain in the muscles of the left calf on walking". His occupation, in a shoe factory, had previously caused him to stand for long hours and during the three previous years he had suffered at irregular intervals from attacks of swelling and pain in the entire left leg. Through these attacks, which were self-limited, he worked. Their exact nature can hardly be surmised. His intermittent limp was such that he could walk no more than a hundred yards without bringing on the cramp-like pain in his calf. There was no radiation. For three months the region of the great toe-nail, especially its outer half, had been dark blue in color. The *cyanosis* faded out upon the base of the great toe and the adjacent foot.

Examination showed a strong, stocky fellow, *suffering* from only a very moderate spontaneous discomfort in his left great toe. The cyanosis has already been described. It was faintly yet unmistakably present on the toes of the right foot as well. The left foot was a little cooler than the right. Investigation with the thermocouple (in the early days of its use) gave very similar temperatures for both feet but there was a very abrupt change just above the left ankle from warm (above) to cold (below). The dorsalis pedis pulsation was absent on the left, faintly present on the right.

Exploration of the left posterior tibial artery behind the internal malleolus, a procedure which might well have resulted in a gangrenous wound, disclosed a shrunken obliterated artery in the midst of a plexus of dilated veins. The wound was made and closed with minute care. The patient was very fortunate indeed that it healed without reaction.

Exploration of the left femoral artery in Scarpa's triangle

terior tibial artery had been obliterated by thrombosis The stump supported an artificial leg for fourteen months At the end of this time a vesicle formed upon its end and though soon healed, gave warning of what was to come, namely, in tractable ulceration

Some two years and a half after first coming under observation, the left leg was amputated through the lower thigh Again healing occurred, again pain was relieved Thrombosis had now occupied the anterior tibial vessels

Six months later, the patient was using crutches, walking on his right leg which had begun to exhibit the disease in the form of a *bursting feeling in the right second toe* However, the Esmarch bandage was now used intensively to secure a reactive hyperemia The flushing time would grow shorter, then longer, but pain was gradually relieved Three years later the right leg was, if anything, better, yet the disease had begun to affect the hands The fingers were numb and cold, especially the little fingers At this time the patient drifted to Mexico and has since been lost to sight

After the first violent attack, the disease in this case seemed to strike rather less strongly, yet its progress, though slower, was never altogether stopped It was peripheral in type, accessions of gangrene coinciding with outbreaks of thrombosis and obliteration of the chief peripheral arteries It would be interesting to know if the patient ever gave up tobacco Doubtless the application of reactive hyperemia saved, for the time being at least, the right leg This is in accordance with general experience, that if an individual can be carried through a bad attack of painful threatened gangrene without a resort to amputation, the collateral circulation which he subsequently establishes by postural exercises and other routine measures will usually save his leg The toes, however, in such a case as this will usually have been left so cold and badly nourished—their nails deformed, their joints stiff—that they are not only useless but obviously threaten further gangrene and sepsis As already explained, their removal, if it can safely be performed, is desirable

in the dorsum of the foot, began to trouble him when he used his clothes-pressing machine. It was this which drove him to seek advice. There was no spontaneous pain.

The patient was a middle-sized, slender man who presented nothing abnormal except for his feet. The blood pressure was 130 mm. systolic. The distal half of the left great toe was deep blue in color, the rest rather less cyanotic but more so than the right toes, which were only slightly discolored. A very distinct moisture of the left foot was taken to be a sign of sympathetic irritation. No pulsations could be detected below the femoral in either leg. Upon cutting off the circulation for five minutes by means of an Esmarch bandage, the reactionary flush advanced rapidly to the base of the toes which did not become colored for twenty to thirty seconds.

Since the signs of disease were of unusually brief duration, the skin in good condition and the patient comparatively young, it was decided to explore the various accessible vessels. The left posterior tibial artery was exposed at the ankle. It was not thrombosed and pulsated very faintly.

The left popliteal artery was exposed by a transverse incision. Here again there was a very feeble arterial pulsation as if the artery were thrombosed a short distance above. The current might of course have been retrograde.

Both these wounds healed well.

Some days later, the left common femoral artery was exposed at the bifurcation of profunda and superficial femoral. The latter was obviously much thickened, as if distended by an embolus, so much so as to be decidedly larger than the common femoral. No pulsation could be detected in the thickened vessel but some blood was evidently passing through it, for, upon being opened, it was found to have the appearance of a rubber sponge. That is, the thrombus which had recently occluded it had been partly organized and canalized. Behind it could be seen several great, soft pulsating branches of the profunda.

Inasmuch as the canalized vessel seemed incapable of carrying a good volume of blood and because a better collateral

was now made The artery proved to be large, soft and very actively pulsating It was not disturbed

Finally, the popliteal artery was exposed It was small, rather thick-walled and, though a feeble current evidently passed through it, did not pulsate A fine bougie passed up into the femoral met an obstruction half way up the thigh Evidently the artery was thrombosed at this point On the principle that the current through the popliteal was feeble and that a better collateral circulation would form if it were divided, a short stretch of it was resected It proved to have been thrombosed and canalized

The result, as is so often the case in arterial resections—corresponding with the experiences of Dean Lewis and Reichert—was favorable The foot became free from discomfort, its color improved and when the patient was dismissed a few weeks afterwards, he was greatly encouraged Two years later, after moving to California, he reported that his improvement had continued, yet his intermittent limp remained Indeed his left became his best extremity, for his right leg was amputated, first below, and later at the knee, and his fingers gave him some trouble Probably he never gave up tobacco Curiously enough, coffee seemed to affect his circulation unfavorably

This case demonstrates, what can now be learned by arteriography, that a good sized artery once thrombosed and canalized is so narrowed as to be incapable of transmitting an effective stream Evidently here the thrombosis occupied both the medium sized arteries of the leg and the superficial femoral

The last case to be quoted is an example of a very mild disease which might have done very well under abstinence from tobacco, which the patient actually gave up, and the postural exercises which were used However, exploration of his vessels in search of information was not only very enlightening but apparently contributed to the rather dramatic result

S B, a Russian born Jew, forty years of age, a cigarette smoker from the tender age of nine, presented himself because of a typical left sided intermittent limp The pain brought on by walking was knife like After two months, a second pain,

Vasomotor signs take the form of unnatural changes in color in response to a cool environment and to emotional upsets. Sweating, which occurs in the abnormal foot while the other is dry, is evidence of an associated sudomotor excitement. A toe which turns blue, then white, then pink again has gone through a cycle of nearly arrested circulation and recovery suggesting vasomotor irritation. And if an individual, subject to vasospasm, is tested by first raising and then lowering his leg, as described in the opening chapter, it will be found that the tips of some toes are very slow indeed to color, remaining white long after the rest of the foot is pink. Yet if a reactionary hyperemia is excited by any of the customary methods, the tips of those same toes will then color like the rest. Vasospasm is likely to be suspected when blueness (or pallor) and coldness of a foot is associated with the presence of a fair arterial pulsation in the dorsalis pedis artery. It should be looked for in excessive cigarette smokers, especially when, with no threat of gangrene, a sudden change to a cold skin-temperature is recognized as the hand passes over the ankle toward the foot.

The final and authoritative test of sympathetic irritability is of course paravertebral blocking of the sympathetic chain with procaine, or, if it is unnecessary to compare the state of two legs, spinal anesthesia. A posterior tibial block with procaine will, if successful, abolish arterial spasm and bring forth a rise of temperature in the toes and sole. A positive is more significant than a negative test. For though the latter suggests an organic constriction, the result may really be due to failure of the procaine to penetrate the nerve.

Vasomotor spasm is important to recognize because it is so often relievable by abstinence [illegible]
and emotional st[illegible]
treatment presen[illegible] described.

Diagnosis.—In the previous chapters, the differential diagnosis between arteriosclerotic vascular deficiency and that of thrombo-angiitis obliterans has been discussed. There the following points were emphasized: as compared with arterio-

circulation would be established if the artery were divided, a block eight cm long was resected

Immediately the toes of the left foot became warmer than the right (lessening of reflex sympathetic irritability?) and within a few days had taken on a normal color In four weeks, under routine treatment by passive vascular exercises and withdrawal of tobacco, the threatened gangrene of the left great toe had cleared up After this, except for the fact that a year later an area of phlebitis migrans was noticed in the left popliteal region, the disease continued to recede Four years later, the patient could walk and even run without a limp The toes of *both* feet continued to be of good color

It is not contended that cases like this, obviously favorably affected by leaving off smoking and the institution of vascular exercises, should be treated by resection of the femoral artery or even exploration of the peripheral vessels The case is put forward as a central, as opposed to peripheral, type of disease, that is, in respect to the limb itself, and to illustrate the vaso dilating effect of resecting a thrombosed artery, a cause of peripheral sympathetic irritation and vasospasm It will be noticed that under routine treatment the early signs of thrombo angiitis obliterans in the opposite leg receded

Vasomotor Manifestations—It has been suggested above that vasospasm is sometimes an element in the ischemia of thrombo-angiitis obliterans and may arise under either of two sets of circumstances first, because such vasomotor nerves as happen to be associated with a stretch of inflamed artery and vein are engulfed in the inflammatory process and, being irritated, excite a vasospasm within the sphere of their influence, both in very small vessels, as in the toes, or in such large vessels as the femoral, and second, because inflammation of a large artery sets up, directly or reflexly, a chronic vasospasm in the vascular tree peripheral to it That this second type of vascular reaction is a real one, however obscure its mechanism, is vouched for by the really dramatic change in the peripheral circulation which often follows resection of a stretch of inflamed thrombosed artery (and perhaps vein)

intermittent limp, in one leg or the other, of coldness in the feet, of difficulty in getting the feet warm at night. Perhaps the great toe and some part of the forefoot on the affected side will tend to become cyanotic and painful on long standing. Pulsation in the arteries of the affected foot will very likely be absent, in the opposite foot, feeble. It may be taken for granted that thrombosis has occurred in the larger arteries below the knee, or even in the femoral above. The prime object of treatment is to encourage the establishment of a collateral circulation.

Smoking must utterly be abandoned. Whether the matter, as Coller and Maddock have shown, is purely one of temporary vasoconstriction with each cigarette or whether tobacco exerts an allergic influence is immaterial. It is agreed today that tobacco smoking, and particularly the inhalation of the cigarette, seriously aggravates the disease and stands in the way of the development of the collateral circulation, the vital widening of the vascular bed. Cutting down the number of smokes is useless and keeps the habitual smoker irritated and dissatisfied. The individual should *know* that he will never smoke again. It will presently be suggested that the pain of impending or actual ulceration or gangrene is more favorably affected by abstinence from smoking than by any other influence. But merely because at an earlier stage of the disease the effect of tobacco is less obvious, is no reason for not giving it up.

Vascular Exercise.—The influence which causes a collateral circulation to increase is, fundamentally, reactionary hyperemia. Just how a generalized vasodilatation of the smaller arteries of a limb shall be secured is immaterial. It is not even certain that for every individual there is one best way. To begin with, vasoconstriction must be avoided. Smoking has already been discussed. Exposure to cold is nearly as harmful. Not only must the feet be protected during the day in cold weather by woolen socks and thick dry shoes but they must be warmed at night by the *warm* water bottle and bed socks. And since cooling the body or even one extremity causes vasoconstriction of the hands and feet, it is about as important to

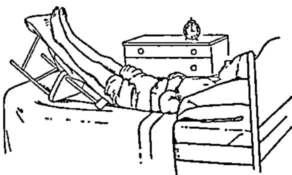
sclerotic disease, thrombo angiitis occurs in youngish individuals between the ages of twenty and forty years, and almost exclusively in males, it is ushered in by intermittent limp, the characteristic reddish blue discoloration of the toes appearing considerably later, it is more given to spontaneous pain and that of a severer character, especially after gangrene has set in, its gangrene is far less extensive, it is sometimes associated with migrating phlebitis, and occasionally it shows itself in the upper limbs

In distinguishing thrombo angiitis obliterans from Raynaud's disease and from the various states of chronic vasomotor spasm, the above criteria are equally useful In none of the states of pure spasm, temporary or permanent, is there intermittent limp, nor is there involvement of both lower limbs in young males Raynaud's disease is a rare disease almost exclusively of females, which shows itself predominantly in the hands The vasomotor changes in thrombo angiitis obliterans unmistakably overlie a disease of chronic vascular deficiency By tests of skin temperature, it will be found that the toes cannot by any sort of reactive hyperemia be warmed to a normal level Indeed, it will seldom be possible to raise their temperature more than a very few degrees as compared with the internal temperature of the body

Treatment

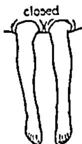
The various stages and forms of the disease require particular treatment, and even those most experienced in its management are not agreed as to the value of certain procedures It is proposed, therefore, to describe, first, the generally accepted method of treating the pregangrenous stage, second, the general plan of treating the disease when gangrene is actually present, and, finally, a number of special forms of treatment which in various hands have proved useful but for which the indications are by no means clear

Treatment of the Pregangrenous Stage—It is hardly correct to speak of such a stage since gangrene need never develop The patient will probably have complained of an

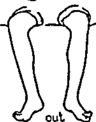


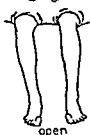












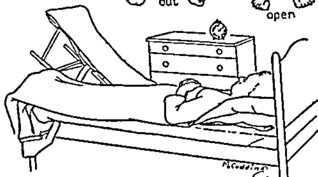


FIGURE 6. BUERGER-ALLEN EXERCISES

protect one part as another By avoiding vasoconstriction, the stage is set for vasodilatation

The Buerger-Allen exercises, carried out as a ritual, are superior to any other measure which does not involve the use of a complicated apparatus The legs are placed successively in three positions

Position 1 Lying on his back, watch in sight, the patient rests his legs upon an inclined plane raised to an angle of 30°–45° He keeps them so raised until the feet are thoroughly blanched, a matter requiring, as a rule, two minutes

Position 2 The legs are lowered and the patient sits with the legs hanging over the edge of the bed, at least, this is the usual direction, but as the side of many beds is higher than the middle and actually offers something like a ridge which presses into the back of the thigh, partly cutting off the circulation, a better position is secured by resting the buttocks against the edge of the bed and letting the legs sprawl out relaxed, the heels resting on the floor However that may be, while the legs hang dependent, the feet and toes are put through a series of motions the ankle is flexed *downward*, then *upward*, the foot is rocked *inward* (tibial flexed) then *outward* (fibular flexed), the toes are *spread* (extended), then *closed* (flexed) As these exercises go on, the feet are becoming flushed They should turn a strong pink, well out upon the tips of the toes, a matter requiring one to three minutes But if they become cyanotic or painful, they should at once be elevated

Position 3 For five minutes the patient lies supine, the legs horizontal in bed and wrapped in a woolen blanket warmed by a hot water bottle or electric pad In this way the reactionary flush, secured by position two, is maintained

The cycle is put through three to six times at one session

The sessions are repeated two to four times each day

A favorable effect is marked by a quicker and more complete flush on depressing the toes after elevation, by a better color of the feet, by an increased range of walking (without exciting the limp) and, rarely, by the return of an absent pulse in the

supply sufficient blood. This is the defect of exposure to dry air overheated under a cradle.

The sitz bath, used at a temperature which feels comfortably hot to the patient is free from danger and usually produces a satisfactory hyperemia—as demonstrated by flushing of the skin. The legs and thighs, that is, the lower half of the body should alone be immersed. A period of ten minutes is sufficient. The sitz bath can be used to start or end the day, even at both times. It comes best, perhaps, after the last exercises of the day, just before going to bed, especially if the patient suffers at all from spontaneous pain.

Diathermy, where suitable apparatus is available, especially when the individual is confined to bed and suffers spontaneous pain, is recommended by some. If it actually conveys heat to the deeper parts, it should do as much as any other agency is capable of doing, and there may perhaps be some advantage in directing heat to some specific locality. However, it is ordinarily sufficient, with a large machine, to place each foot upon a metal electrode in order to route the current through both legs With a small machine, diathermy can be applied to one leg or a part of one. The amount of current must be gauged by the patient's reaction, his sensation of warmth and comfort. For ambulatory patients treatments every other day may be sufficient. Bed patients can be treated daily.

Vascular Exercise by Special Methods more elaborate than the Buerger-Allen exercises include (1) the Oscillating Bed, (2) the Suction and Pressure Boot and (3) Intermittent Venous Occlusion.

The Oscillating Bed is an expensive labor-saving device for doing Buerger's exercises. The patient need make no exertion and the exercises can be continued day and night. Whether the use of the bed entails any disadvantages, as for instance in the direction of a bad effect on the thoracic organs or brain, does not seem to be known. It certainly is not an essential means of treatment.

The Suction and Pressure Boot has not been particularly useful in the treatment of thrombo-angiitis obliterans. Not

dorsalis pedis artery Such spontaneous pain as may be present is apt to be relieved Except for the favorable reaction to abstinence from smoking, which is apt to be rapid, the improvement is usually gradual So far as active exercise is concerned, walking should not be attempted except within the limits of comfort, that is, the intermittent limp should not be excited The value of exercise lies of course in overcoming muscular atrophy and tends to break up the vicious circle of atrophy and a diminished vascularity of the muscles

A judicious mixing of rest and moderate regular outdoor exercise should tend to raise the low blood pressure which a considerable number of cases present Raised pressure forces blood more abundantly through narrowed vessels and opens up fresh collateral channels Drugs, except in so far as they may improve the appetite, diminish fatigue, or confer a sense of well being, are of no advantage

In addition, the careful toilet of the toe nails, toes, and feet, as prescribed for arteriosclerotic and diabetic states, should be put into effect the feet and toes dried with a soft towel after the morning wash with warm water, light greasing with a cold cream or lanolin, softening and filing of deformed nails, exquisite care of corns and calluses, and, finally, treatment of any fungus infection which may be proved to be or may even only be suspected of being present

It goes without saying that the blood should be studied for any gross abnormality—no consistent chemical changes encouraging thrombosis have been discovered—and that any such should be corrected, that the fluids should be kept up by an abundant intake of water, and that any discoverable infectious foci should, on general principles, be abolished

The Application of Heat —Whether heat should be applied depends somewhat upon the patient's reaction to the routine already described, for the reactive hyperemia and the avoidance of vasoconstriction can of course be secured without recourse to specific means of heating the limbs It has already been explained that too high a degree of heat sets going a metabolic activity for which the circulation is often unable to

supply sufficient blood. This is the defect of exposure to dry air overheated under a cradle.

The sitz bath, used at a temperature which feels comfortably hot to the patient is free from danger and usually produces a satisfactory hyperemia—as demonstrated by flushing of the skin. The legs and thighs, that is, the lower half of the body should alone be immersed. A period of ten minutes is sufficient. The sitz bath can be used to start or end the day, even at both times. It comes best, perhaps, after the last exercises of the day, just before going to bed, especially if the patient suffers at all from spontaneous pain.

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uncommonly, even in the production of negative pressures of sixty to eighty mm of mercury, the heavy rubber cuff which surrounds the thigh seems actually to check the arterial inflow This is particularly true in the advanced case In mild cases a reactive hyperemia and a collateral circulation can be brought out by less elaborate and expensive methods In the presence of gangrene, the use of the boot leads to a spread of infection However, it will be worth while to try the apparatus on many cases in which spontaneous pain without gangrene is a feature In unpredictable instances, pain is certainly relieved It will *not* be worth while to make financial sacrifices to secure the use of an apparatus which can accomplish little not equally obtainable by simpler means

Intermittent Venous Occlusion —Any one possessing a blood pressure machine can carry out this treatment, though for routine hospital use the apparatus of Collens and Wilensky is labor saving and capable of repeating the occlusion with whatever pressure is desired For persons with low blood pressure and a deficient peripheral circulation, the compression, which in any case should not be higher than the diastolic arterial pressure, should range perhaps from forty to sixty mm of mercury and should not be maintained for more than two minutes at a time A broad cuff for the thigh is desirable De Takats has pointed out the advantage of elevating the leg for a minute or so after each compression—to drain it of blood—and the disadvantage of continuing the exercise for bouts of longer than one half hour

Intermittent venous occlusion should prove more useful in the treatment of thrombo angiitis obliterans than suction and pressure in the boot, for, as already explained, it produces pretty much the same physiological effects but without unduly compressing the feeble arterial stream It certainly requires less expensive apparatus and is easier to adapt to home use

Intravenous Saline Injections —It is very difficult to know what to say about this particular treatment The suggestion of diminishing the blood's viscosity came from Japan Willy Meyer introduced it into this country, and such experienced

clinicians and investigators as Samuels and Silbert swear by it. Originally, a physiological salt solution was used, sodium citrate solution has been tried, and after considerable experience with hypertonic saline at a strength of five per cent, the tendency at present is to use a two or three per cent solution of sodium chloride.

There is no question that at one time rubber tubing and salt solution insufficiently freed from foreign material—in other words, dirt—had a good deal to do with the favorable effects the patients experienced or thought they experienced. They were, in fact, subjected to protein shock with its accompanying chills and fever. Doubtless this aided vasodilatation. Yet it seems to be established, by oscillometric tracings as well as by clinical experience, that the repeated intravenous injection of *slightly* hypertonic clean salt solution enlarges the amplitude of the peripheral pulse. The effect of any one injection may last several hours. The patient is said to notice first a feeling of increased warmth in the affected leg. Trophic disorders tend to clear up. Above all, the intermittent limp is said to improve so that the patient can walk farther and faster. A decided advantage of the treatment is that it can be used in the presence of gangrene and ulceration when passive exercises and intermittent suction or venous compression may be harmful. Also it offers the patient a routine whose potential usefulness he can see and feel, an important consideration as those will testify who have had to see patients through a long bout of gangrene and agonizing pain. Its disadvantage is the nuisance of repeated [illegible]
salt solution and tubing, [illegible]
well-equipped hospitals.

[illegible]
tior [illegible]
to [illegible]
[illegible] water. After filtering into a 500 ccm. pyrex glass flask, the solution is either sterilized in a pressure autoclave for ten minutes or boiled vigorously for the same time. The extra fifty ccm. are to allow for evaporation. For a two per cent solution six grams of salt are used.

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The surroundings should be as cheerful and diverting as possible. An abundant fluid intake should be maintained. The leg should be kept warm by woolen coverings. If the toes are not actually gangrenous or ulcerated, Buerger-Allen exercises may be used, or intermittent venous occlusion may be tried, or alternating suction and pressure in the boot. Heat can be applied in the form of diathermy, or a properly controlled electric pad (if there is such a thing) outside the woolen wrappings. In other words, if ulceration and gangrene are absent, all means of exciting a reactive hyperemia will be tried, with the expectation that pain, if present, will be relieved as the peripheral circulation is improved.

In the presence of ulceration and gangrene, vascular exercise will have to be confined to intermittent venous occlusion (arterial occlusion may cause additional thrombosis and though useful if successful, is a dangerous gamble) and even the occluding venous pressure must be light—hardly more than forty mm. of mercury. Under these conditions, a trial of two or three per cent saline solution, to be injected in an amount of 300 ccm. three times a week is certainly worth considering. Diathermy can still perhaps be used. To assist sleep and assuage pain, alcoholic drinks will be of some help as well as offering aid in vasodilatation. For drugs, the barbiturates will have to be used, morphine never, though codeine sulphate may perhaps be combined with the other sedatives.

The local treatment of an ulcerated area, as for instance the common form beside the margin of a nail, may properly consist in a daily short soak in a warm solution of almost any mildly antiseptic sort. This is to loosen adherent secretion and favor drainage. A freshly prepared solution of chloramine, 1-200 is satisfactory. The watery solution of iodine in the form of Lugol's solution 400 times diluted is not irritating. A watery solution of a cocoanut oil derivative; anything clean will do, including a saturated boracic solution or normal saline provided it is sterile. Then the sore should gently be dried with cotton swabs, the adjacent skin cleaned with cotton and a neutral soap, wiped off with the same antiseptic already

The original solution is resterilized eight hours later for an other ten minute period When a needle of nineteen gauge is used, the injection should take about ten minutes Three injections a week are advised, for a three months' period, and are then gradually discontinued during the next six months For patients whose oscillometric index is zero or less than 0 5 at the ankle, a longer period of treatment is recommended

Diet and Drugs —A diet high in calcium and low in potassium has been suggested, apparently because of its favorable affect on vasospasm Perhaps it is more important merely to see that the patient has an abundant, *attractive,* varied diet, with the idea of keeping up his nutrition during the difficult stage of the disease Three good meals a day are something to look forward to during an otherwise boring if not actually painful few weeks or months in bed Vasodilating drugs can not be recommended

Sedatives and hypnotics are a problem Opium and its derivatives are absolutely barred If effective, they set up a habit, but actually they soon lose their effect The routine measures already described must be relied upon to subdue pain, but for sleep the barbiturates are probably the most useful From the great number of preparations available some few will usually be found to be satisfactory

The Treatment of Threatened or Actual Gangrene —Patients showing deep cyanosis in one or more toes, or ulceration, or actual gangrene, usually suffer also from spontaneous pain The vicious circle of edema, a defective arterial supply and a preference for the dependent position (to relieve pain) is therefore almost certain to be present Smoking is first of all barred The patient is confined to bed and if possible to a bed whose parts can be tilted so that the foot can be raised for a time or lowered (This does not refer to the automatically tilting beds earlier mentioned) The plan is to keep the affected leg at least horizontal for nearly the whole twenty four hours if possible—to get rid of the edema—and only when the patient can bear elevation no longer to secure relief by the least possible amount of depression

He may even employ paravertebral sympathetic resection. The indications for all such special procedures are not clear. Only a considerable familiarity with the ins and outs of thrombo-angiitis obliterans and of the procedures themselves justify their use on special occasions.

Surgical Treatment

Peripheral Nerve Section.—Though frowned upon by many and requiring a high degree of technical skill and judgment, section of the sensory nerves supplying the sole and toes may offer the only alternative to a major amputation in the presence of uncontrollably painful gangrene. Actually, since the importance of cigarette smoking has been recognized and abstinence from tobacco has been enforced, the very painful gangrenes are seen far less often than formerly. It may be possible, therefore, to carry the individuals suffering from the more painful types of disease through their bad weeks or months without resorting to nerve section. Two procedures are available: (1) blocking the sensory branches of the superficial and deep peroneal nerves just below the head of the fibula and (2) blocking the superficial peroneal, anterior tibial and posterior tibial nerves in the lower middle third of the leg.

Blocking the superficial and deep peroneal nerves at a high level.—This procedure is done through a three- to four-inch incision which begins just above and medial to the head of the fibula and is carried straight downwards. If carefully handled, the wound is almost certain to heal. The common peroneal nerve is found dividing in the substance of the extensor digitorum longus muscle, which is extensively split. The important motor branches come off within two inches of the head of the fibula. The sensory branches can be identified electrically but this is hardly essential because even if a partial foot drop results from crushing the supposedly sensory branches (as happens in perhaps fifty per cent of the cases) the benefit of the procedure is still greater than its demerits, and as the nerves recover, the moderate degree of paralysis disappears. The sensory nerves are best crushed with a hemostat for per-

used, and the area next to the ulcerated or gangrenous surface covered with vaseline gauze An actually gangrenous part can be treated like an ulcer unless the gangrene is already so dry and the area nearby so clean that anything but a sterile dry dressing would be out of the question

To aid in controlling pain, an analgesic drug may be added to an antiseptic ointment Such may be successful if an ulcerated surface is present but can have little or no effect on normal skin or gangrenous tissue Ethylaminobenzoate ointment, ten per cent, or nupercaine, one per cent in petrolatum, may be useful

When a large open surface is left, as after the casting off of the end of a toe, the exposure of a joint or the formation of a crevice between the toes, the use of Dakin's solution may prove possible The full ritual must be used, with protection of the skin by vaseline gauze The oily preparations of the hypochlorites are often preferable, being less painful, as for instance dichloramine T in chlorcosane, or eusol and mineral oil freshly mixed in equal parts Combinations of zinc oxide, mineral oil and cod liver oil promote healing and epithelization

Should a good line of demarcation form at the base of a toe, with dry gangrene peripherally and reddened reactive skin proximally, amputation in the zone of reaction is permissible, but such a state as this will seldom present the problem of painful gangrene for which treatment is so difficult In other words, local amputation, even if surgically possible, will seldom relieve the pain of gangrene And when, as a result of the casting off of gangrenous tissue and the appearance of healthy granulations, the tissues are ready for healing, the battle is won anyway In the presence of signs indicating this outcome, conservative treatment is continued, however slow progress may be

In aid of such treatment as the above, the resourceful surgeon may invoke the assistance of some of the procedures described in the following paragraphs, as for instance, sensory nerve block, periarterial sympathectomy, or arterial resection

sympathetic supply to a limb by removal of the ganglionated chain in the lumbar region, or dividing the preganglionic fibers in the upper thoracic, may be counted a last resort in the treatment of thrombo-angiitis obliterans. If the routine measures—abstinence from tobacco, rest, protection from cold, a liberal diet, and passive vascular exercises—fail to cause improvement, the procedure may be considered. But before accepting the operation, one must go much further. If results of any consequence are to be expected, it must be shown, by the trial of some of the tests of reactionary hyperemia described in the opening chapter—brief arterial occlusion, heating the other limbs or the body, the intravenous injection of sodium nitrate or, best of all, spinal or paravertebral sympathetic block—that the peripheral cutaneous temperature can be decidedly raised or the peripheral pulsations increased in the limb in question. Using these criteria, sympathetic resections will not often be performed. However, in individuals reasonably young, as sufferers from this disease must usually be, and in persons whose general health is reasonably good, the operations present no especial difficulty. For the lumbar sympathectomy, an extra-peritoneal approach through the flank, leading to removal of the second and third lumbar ganglia, is satisfactory. For the upper thoracic operation, there are two approaches,* from above the clavicle and from the back. In case of threatened loss of the fingers, it is believed that the upper thoracic resection is usually worth trying. That an alcoholic injection, as described in the previous chapter, will serve the [illegible]ose [illegible]nd [illegible] of [illegible] channels.

In case of threatened loss of a leg, the lumbar sympathectomy is of doubtful value. If pain, plus gangrene, for which amputation is usually performed, is not relieved by other means, lumbar sympathetic resection is also likely to fail.

As a cure for intermittent limp, lumbar sympathectomy is not recommended.

* These operations are fully described in the following chapter.

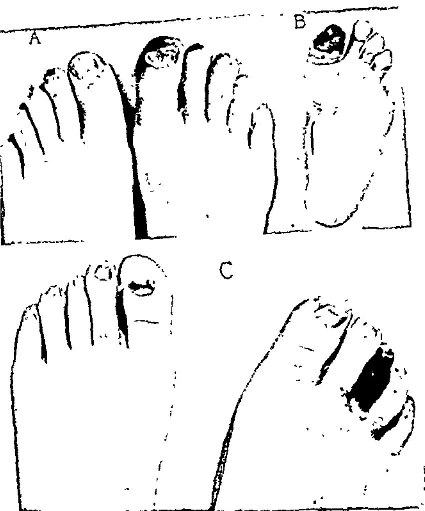
haps 0 5 to 1 cm Regeneration does not recur until the patient has been tided over the difficult period

Blocking the superficial peroneal, anterior tibial, and posterior tibial nerves —This operation, as devised by Smithwick and White, is performed rather below the middle of the leg but not below a point five inches above the ankle joint Plate IIIB, page 107, shows the anatomy of the nerves and the distribution of the cutaneous branches The posterior tibial supplies the sole so that crushing this nerve leaves that part, together with the tips of the toes, anesthetic and paralyzes as well the intrinsic muscles of the foot The only advantage of the lower incisions is to avoid the possibility of toe drop The wounds must be made with great care lest sloughing occur Smithwick and White advise making the incision to reach the anterior tibial midway between the tibia and fibula. The superficial peroneal can be picked up through this same incision For the posterior tibial, the incision is parallel to the posterior angle of the tibia The cleavage plane between the flexor digitorum longus and the soleus gastrocnemius muscle is the guide to the nerve The nerves are to be crushed with the hemostat

The injection of alcohol into these nerves too often results in an alcoholic neuritis causing pain nearly as bad as the original But there is no objection to dividing the nerves as Silbert has suggested Indeed Silbert was the first to attempt the relief of pain, using alcohol but without exposing the individual nerves

The benefits of blocking the sensory nerves go rather further than the mere relief of pain The dressings become very much easier, and whereas the forefoot may have been so sensitive that Dakinization could not be used, now the application of Dakin's fluid or other hypochlorites is possible Thus gangrene and ulceration may rapidly be cleared up, and sources of pain eliminated before the nerves can regenerate Moreover, the peripheral blood supply may actually be improved (release of vasoconstriction)

Sympathetic Neurectomy "Sympathectomy"—Blocking the



EARLY THROMBO-ANGIITIS OBLITERANS *A* P B., aged thirty, a heavy smoker Mass Gen Hosp 327844 Early, minor gangrene of several toes, probably associated with severe fungus infection Local treatment and lumbar sympathectomy saved toes *B* J A R., aged thirty-eight, a heavy smoker Univ of Virginia Hospital Local gangrene, well demarcated Little toe recently amputated Remaining toes healthy and limber Prognosis good *C* H P., aged twenty four Mass Gen Hosp 317313, a heavy smoker Intermittent limp for one year Local gangrene beneath *left* great toe-nail two weeks earlier—healed *Right* foot, sudden agonizing pain four weeks earlier. Note shiny dark (red) skin and gangrene of third and great toe Rapid advance of gangrene—amputation through thigh Almost certainly, sudden occlusion of a large peripheral artery by thrombosis

PLATE II

Arterial Resection Periarterial Sympathectomy—In some respects, these two procedures may be expected to act in a very similar way Both may break up a vicious circle of local vascular irritation and peripheral vasospasm An example of the good effect of resecting the femoral artery in an instance of femoral thrombosis has been quoted earlier in this chapter An extensive femoral perivascular sympathetic extirpation acts perhaps in a somewhat similar, if less radical way (It fails of course to divert the stream from a partly closed artery) But the indications for either procedure are not well established Neither should be used by any but those most familiar with the disease

Amputation—The operations employed are those already described in the treatment of arteriosclerotic disease, but the criteria are somewhat different For most individuals will still desire to be active and, if kept active, will be able to earn their own living The danger of infection is decidedly less Therefore if the routine observations and tests show a reasonable vascularity, giving hope that an amputation *below* the knee will result in a useful stump, the amputation through the leg should be tried There is all the more reason to save the knee joint since a disease severe enough to cause the loss of one leg will often involve the other to a similar degree, and one natural knee joint is a tremendous advantage, two artificial knee joints being too great a handicap for any but the most courageous and athletic

The amputations through the knee joint* of the Gritti Stokes or Callander type will of course be most generally useful, and a higher amputation is seldom necessary

There is no doubt that amputations today are resorted to much less frequently than before the significance of tobacco was generally accepted The fearfully painful intractable forms of the disease are now less common and doubtless in the future will be rare In the treatment of thrombo angiitis obliterans, amputation of more than the toes is decidedly a confession of failure

* These operations were briefly described in the previous chapter

Thrombo-angiitis Obliterans in Women

The rarity of the disease among women who, until recent years, have not been cigarette smokers, has always seemed somewhat of an argument that smoking is an important factor which at least aggravates thrombo-angiitis obliterans. In 1932, Horton and Brown, of the Mayo Clinic, were able to collect only ten cases in females, out of some seven hundred instances of the disease which they felt might properly be counted thrombo-angiitis obliterans. The average age of the women was thirty-nine, six of them presented characteristic peripheral lesions, and two had suffered from a migrating phlebitis. Curiously enough only two gave a history of intermittent limp. The disease on the whole seemed milder than that of men. Many do not believe that it exists.

The following case is an example of a severe form of the disease * observed at the Peter Bent Brigham Hospital.

S.L., a married Polish woman, forty-five years of age, the mother of one child, desired to smoke cigarettes so continuously that although hospitalized during November, December, and January, she was kept outdoors for the greater part of each twenty-four hours.

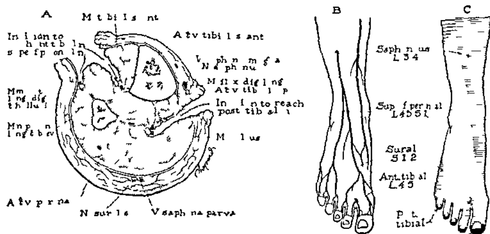
Since the age of fifteen, she had been troubled by an intermittent limp, worse in the right leg than the left. Seven years before first entering the hospital, at the age of thirty-seven, she began to suffer from spontaneous pain in the right foot and knee. A white patch appeared upon the right foot which became very cold and as if needles were being applied to it. Rather suddenly, then, the right foot turned purple and the leg was amputated through the right thigh.

While still convalescent from the amputation, the left foot

* In a letter some years later, the patient recalls her habit as a little girl of eating ergot in the form of the fungus as it grew on the rye In her own words "When I was very young I used to walk through rye fields on my way to Church, during the month of June just before the rye ripens *** Some of the kernels will turn black and grow to the [illegible] torted kernels and eat them as [illegible] them. Many a time I picked a [illegible] Possibly her disease is in fact e[illegible]



1 Gangrene of the Most Vicious Type, in a Female S L, aged forty five, a very heavy cigarette smoker Diagnosis of Thrombo angiitis Obliterans made upon amputated leg The other leg had already been amputated several years earlier History of eating ergot



B Distribution of the Sensory Nerve Supply to the Foot Showing White and Smithwick's method of exposing the various nerves about five inches above the ankle Crushing them at this level does not cause toe drop, but leaves the sole anesthetic and paralyzes the intrinsic muscles of the foot (From Homans' *Textbook of Surgery* Courtesy of C C Thomas, Springfield, Ill, and Baltimore, Md)

PLATE III

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became cold and swelled at the ankle Nevertheless, except for a sense of coldness and "needle pricking" in the left foot, she remained active, using a right artificial limb successfully Heat brought relief of the symptoms of discomfort in the left foot

One month before entering the hospital, the patient noticed, for the first time, a whitish area upon the dorsum of the left great toe In two weeks, this area, which the patient had con cealed with a bandage, discharged pus Soon after this, the great toe became gangrenous, the fourth followed suit and an area of gangrene spread across the forefoot At the same time spontaneous pain became fixed and fairly severe Hang ing down the foot brought some relief

The state of the foot is sufficiently revealed by the photo graph reproduced It will be noticed that the area of gan grene is irregular and that the foot exhibits a moderate edema No pulses could be felt in any vessel below the inguinal liga ment

During the next six weeks the gangrene advanced moder ately, always having a well marked line of demarcation Pain was rather intermittent, occurring chiefly in a crampy form at night The cutaneous temperatures of the foot and leg were not particularly low There was no difference between that of the thigh and leg No abrupt change anywhere The mouth temperature ranged daily from 99° to 100° F Spinal anes thesia caused no vasodilatation whatever in the left foot The patient could not be prevented from smoking constantly!

Amputation, which, in view of the loss of the other leg above the knee, might well have been attempted below the knee, was made through the lower third of the thigh and healed after only minor sepsis confined to the skin The patho logical report was "Thrombo angiitis obliterans" and the description of the diseased tissue is consistent with the ap pearances in authentic cases among men

Two years later the patient expressed herself as being well and cheerful She was successfully navigating a wheel chair and smoking as much as ever

CHAPTER IV

SPASM OF THE ARTERIES AND ARTERIAL EMBOLISM

Maurice Raynaud gave the first systematic description of a disease characterized by arterial spasm. As is usual in the observation of a new symptom-complex, he included in his account of thirty cases a greater variety than he supposed. It has even been maintained that among the thirty there is only one instance of Raynaud's disease! In which case, posterity would seem to have been rather fussy and ungracious, for Raynaud knew enough about recurrent vasospasm to give a description which has hardly since been bettered. His fault, if any, lay in trying to explain, as a single phenomenon, too great a variety of conditions. Naturally he began with "dead finger". "Madame X", said he, "had been subject since childhood to an infirmity which makes her an object of curiosity to her acquaintances". Any sudden cooling of the atmosphere, even in summer, would cause her fingers to become bloodless, without feeling, and of a whitish-yellow color. She would wring them violently or soak them in lukewarm water; whereupon the vascular spasm would gradually relax, to be succeeded by a very painful reaction. That this was a neurosis was suggested by the prompt disappearance of the attacks with the onset of pregnancy.

Raynaud ... young wo ... of such pe ... chilblains, in fact, the unnatural reaction to cold, so common in females and by no means unknown in males, in the damp winter climate of western Europe and of England. However, he reasoned correctly that ...

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Raynaud noted the frequency of this sort of thing in young women, especially in times of damp cold, the tendency of such persons to chilblains, in fact, the unnatural reaction to cold, so common in females and by *no* means unknown in males, in the damp winter climate of western Europe and of England. However, he reasoned correctly that this was only a mild exhibition of a very serious process which could and did lead to nutritional changes in the form of actual necrosis of the

finger tips, and to a lesser degree, of the toes It is the easily excited, recurrent, vicious vasospasm, causing minute, oft repeated necrosis with conical deformation of the finger tips, which bears Raynaud's name today, a rare disease But Raynaud is responsible for very much more than this, for since his time, clinicians have been on the lookout for many other sorts of vasoconstriction—acute, recurrent, and even chronic arterial spasms which discolor fingers and toes, hands and feet—painful states as a rule and very often marked by organic changes, ranging from small necroses and ulcers to gangrene of one or more digits It has been learned that acute arterial spasm can arise from trauma to a large vessel or even to the limb it supplies, and from injuries to great nerves such as the median or sciatic—causalgias and causalgia like states—resulting in painful ischemia of the peripheral parts, that an arterial embolus not only plugs a great vessel but sets up a widespread vasospasm in the vascular tree beyond, and even that thrombosis in an important vein may excite such spasm in the companion artery as to cause gangrene of an extremity Still more obscure and peculiar are the vasoconstrictions associated with old infantile paralysis and other nervous disorders

In the unravelling of these various symptom complexes the names of Sir Thomas Lewis and Rene Leriche stand out, the first for his sound physiological investigations, the latter, for his brilliant reasoning and surgical feats But many peculiar states remain to be identified and explained, and so the writer makes no apology for presenting the subject of vasospasm with little attempt at order and for including, it may be, in one category conditions which, for all that they are related to spasm of arteries, large or small, may well be due to a considerable variety of causes After all, the main thing is to recognize in any one case, that vasospasm is or is not the hand behind the disturbance and, having decided in the affirmative, oppose its action as effectively as possible

At this point it will be well to review the account of the sympathetic system more fully given in the first chapter Normally, the outflowing sympathetic influences distributed to the

blood vessels of the entire body maintain an even arterial tone. The nerves controlling the muscular arteries are distributed from the spinal cord through preganglionic fibers to the sympathetic gangliated chain, which lies close beside the vertebral column. Thence, postganglionic neurons pass out with the great nerves of the limbs to ramify at intervals over the larger arteries (Figure 2), and finally, just as sensory nerves are distributed to the skin, so the vasomotor fibers are assigned to the fine arteries and arterioles of corresponding peripheral fields. Thus a general stimulation of the sympathetic system causes vasoconstriction over the entire body; or a stimulus touching only the sympathetic supply to one arm excites vasospasm in that arm. The disorder may even be so local as to affect the vasomotor fibers distributed through a single nerve. Sudomotor and pilomotor excitement is associated with vasoconstriction. A feature of the sympathetic system in the hands and feet is the very rich supply of fine arteriovenous connections capable of being closed or widely opened according as the sympathetic contracts or relaxes their walls. By means of these connections, the surface of those terminal parts conserves heat or gives it out, and thus arterial spasm shows itself more plainly in the hands and feet than elsewhere, particularly in the fingers and toes. The coldness and pallor it occasions are almost necessarily associated with sweating. On the other hand, the arterial dilatation of sympathetic paralysis leaves the skin of the extremities hot, pink, and dry. The variations upon these reactions will be brought out in the account of the various vasospastic states which follows.

Raynaud's Phenomenon

This title is intended to include all recurrent arterial constrictions of the extremities which are excited by cold and the emotions. Such vasospasms are common. It is only the serious, progressive, vicious spasms, which Raynaud himself accused of causing nutritional changes, that are rare. These latter will be described as Raynaud's disease, for that, whatever Ray-

naud himself may have written, is the accepted nomenclature of today With this understanding, the comparatively mild and common form will be distinguished as Intermittent Spasm of the Digital Arteries, a Reaction to Cold Thus all members of the Raynaud family will be housed under one roof, which, after all, is as it should be, for most of its members appear to be *sisters*

Intermittent Spasm of the Digital Arteries, a Reaction to Cold—The disease, if so it can be called starts in childhood or adolescence, rarely later, and is decidedly more common among girls than boys, though there is a familial form, picturesquely described by Hunt in his *Critical Review of the Raynaud Phenomenon*, as "Hereditary Cold Fingers" which appears in both sexes There are no associated organic peculiarities of the nervous system or in the body at large The hands rather than the feet are affected, the fingers rather than the thumbs Symmetry is the rule, even in the unusual event that only one finger exhibits the disease, for instance, the middle finger of each hand may alone be sensitive to cold The subjects are apt to be thin and, in the countries where the winter cold is *damp*, are likely to suffer from chilblains Thus, digital spasm, and for that matter progressive Raynaud's disease, is far more common in England and Europe than America

The attacks, which often start in childhood, are brought on by exposure to cold, a cold atmosphere, as a rule, during the winter months, but equally well by bathing in cold water Sometimes, if cold alone has failed to produce an attack, added excitement or embarrassment will do so There is a great variation in the susceptibility of individuals Some react to the slightest sort of exposure Some will only notice whiteness and numbness of a finger after being in swimming for hours, while others will find that all the fingers are involved if they poke their noses outdoors on a cold day Sometimes the tips of the fingers only are affected Lewis has called attention to the fact that when the attacks are provoked by placing the hands in cold water, there is a definitely optimal

temperature. For instance, water at 15° C. (59° F.) brings on a spasm, while ice water turns the fingers rather red than white. The first sign of local vasoconstriction is pallor of the finger-tips. The pale area soon turns gray as the remaining blood loses its oxygen, but this initial change may be inconspicuous. When the individual first notices her fingers, the tips or even a considerable portion of each have usually become waxy white. Actually these changes depend a good deal upon the original color of the skin. A rather highly colored skin turns bluish and fingers already pale become waxy. Moreover, if the spasm relaxes at moments, fresh blood flows in and soon, giving up its oxygen, makes the skin cyanotic.

The course of any attack will depend upon the length and severity of the exposure. If the fingers are made white and bloodless for long, a numbness sets in at the tips and in half an hour may involve the whole of each digit. Particularly severe attacks are very disagreeable. After a while, as the hands are warmed and spasm is relaxed, a slow wave of bright redness passes out upon the fingers, a reactionary hyperemia, as after any arterial stoppage. Under this influence, each finger is apt to tingle, becoming warm and slightly swollen. A capillary pulse can often be detected at the finger-tip. Such a reaction, as Raynaud noted, may be painful.

The course of this mild disease is rather favorable than otherwise. Rarely it dies out as the years advance. Or the individual learns to avoid the combination of events which excites the attack. Perhaps the spasm appears so seldom that it can be ignored and repeats itself, rather to the victim's amusement, on occasion. Should the attacks become progressively more severe, the disorder must be placed in the class presently to be described as Raynaud's disease.

Treatment.—Madame X did about as well as anyone can. She wrung her hands and placed them in lukewarm water. Whether pregnancy relieves others as well as her seems not to be known, so that as a form of treatment it can not be accounted reliable! But of course prevention is really more important. There are, as Lewis points out, two elements in

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well to emotional (adrenal) influences. Here the matter may properly be left, with the understanding that the arterial spasm of Raynaud's disease occurs in the digital arteries, the nature of the local fault being obscure. It is only necessary to add that in advanced Raynaud's disease the walls of the digital arteries are actually thickened, so that their capacity is decreased. Under these circumstances, *any* influence which increases vasoconstriction—cold, embarrassment, anger—will be able to close them altogether. Ross has been able to determine the amount of structural change in the digital arteries of persons suffering from Raynaud's disease by the speed and amount of the rise of cutaneous temperature in response to warming the body. In mild, early cases, the arteries are capable of full dilatation. In advanced cases, vasodilatation is very slow and incomplete.

The same sort of individuals—almost always women of youthful years, though the disease occasionally appears in middle age—as are susceptible to the milder form of Raynaud's phenomenon suffer from serious progressive Raynaud's disease. The attacks are now more easily provoked and more prolonged. They start in the finger-tips and mount to the base of the fingers, sometimes to the palm, but almost never above the wrist. The first color is usually a bluish one which deepens to slate blue or a dark purple. If the attack is prolonged, the cyanosis is replaced by a waxy pallor. In any case, no return toward a normal color can occur until the hands have been thoroughly warmed. Partial relief is marked by a shift from blue to red as a little arterial blood pushes into the fingers. Indeed, this betwixt-and-between state may last for some time, the red and blue areas existing in a patchy way; or perhaps a whole finger will shift back and forth, never really becoming warm and pink. Sooner or later, as the attacks multiply, the pulp of the fingers hardens, the skin tightens, and its transverse wrinkles disappear. The fingers taper toward their ends, the overcurved nails projecting beyond the shrunken tips. Upon these tips, little necrotic areas appear in the form of tiny cores of dead skin, which separate

this sort of vasospasm There is the local spasm which results from direct exposure of the fingers and hand to cold, and there also is the vasoconstriction, in which all the peripheral circulation shares, due to lowering of the body temperature as a whole Thus the individual must be on the lookout against immersing the hands in cold water, or exposing them without woolen coverings in cold weather, or letting the fingers come in contact with cold metal or glass But she must also avoid chilling the body or, in case the disease shows itself only in the hands, any other part, especially the feet It may be well, as is true of those suffering from the serious progressive form of the disease, to begin the day by heating the hands in hot water, establishing a vasodilatation which will not easily re act to slight cooling

Raynaud's Disease (Raynaud's Phenomenon with Nutri tional Changes) —Lewis's experiments in exciting spasm in the digital arteries by exposure of the fingers, even the base of the fingers alone, to cold, and his demonstration that such spasm cannot be excited by any central sympathetic stimula tion, provided the hands are kept warm, strongly indicate that the fault in Raynaud's disease is a local one If this were all, the whole trouble would seem to lie in the state of the digital arteries themselves This conclusion, however, cannot unre- servedly be accepted It has been shown that vasospasm in the fingers can take place, especially under the influence of emotion, when the peripheral portion of the sympathetic is absent Indeed, as Smithwick, Freeman, and White have dem onstrated, removal of the ganglia whose cells supply, through the gray, or postganglionic, rami, vasomotor fibers to the pe ripheral arterial tree, leaves such arteries exposed to adrenal influences, under which peripheral spasm, as excited through the adrenal secretion by any emotion, may be exceptionally severe Moreover, Smithwick, Telford, and others have found that if they divide the *pre*ganglionic rami of persons suffering from Raynaud's disease, leaving the peripheral neurons in tact, these persons appear to be cured of their old tendency to peripheral spasm in response to cold and to be resistant as

passed then to a violet tint, then to a slaty white. * * * A very cold wind had blown all morning when Rose entered the room. Her cheeks and chin were of indigo colour; her hands were as cold as marble. At the first view I believed them to be gangrenous. The ends of the fingers were of a greenish blue, the palms of the hands were of a deep purple. On the forearms there were marblings similar to those which are present on the legs of persons who use foot warmers. Above the wrists the skin presented its natural colour. These phenomena were less pronounced in the lower limbs, which were clothed with woollen stockings. * * * Whilst this young woman spoke, a bright redness began to develop itself at the root of the nose and over the cheeks; then it extended and invaded the blue colour, which soon formed no more than a deep red patch on the tip of the nose, and ended by disappearing entirely. One moment afterwards, the pink colour of the nose began to pale, and this organ resumed little by little its ordinary colour. The same change came into operation at the same time upon the cheeks and the skin; upon the hands the change did not take place so quickly nor in the same way. * * * It was at the extremities of the fingers that the cyanosis and the cold persisted longest. Finally at the end of a quarter or half an hour the whole hand was of vermilion red; the pulse had regained its force, the warmth of skin was perfectly developed, and a slight sweat had moistened the cutaneous surface. All these phenomena were reproduced each time that Rose was exposed to cool air, whether in the evening, morning, or at the middle of the day. The reaction only commenced when she returned to her room."

On the whole, the appearance of gangrene away from the extremities and in such parts as the tip of the nose, the cheeks or the pinna of the ear, that is, a juvenile gangrene, is less likely to be caused by a Raynaud-like vasospasm than by minute multiple arterial embolism (see page 162). Nor are the contractions of the retinal arteries, which Raynaud observed in one of his later cases, at all characteristic of the typical disease. Vascular spasms, it is now realized, are ex-

painfully, leaving minute scars The process shows no sign of gross ulceration or infection, by contrast with the outspoken gangrene of arteriosclerosis or thrombo angiitis obliterans, but is painful, sometimes agonizing Moreover, so far as the limbs are concerned, it begins in, and is decidedly most advanced in, the fingers rather than the toes But even if the loss of substance is slight, pain is out of all proportion to it, and healing of the little areas left exposed by the casting off of the tiny necrotic plugs is very slow However, even in the severest form of the disease, years go by before the fingers are noticeably shortened

Manifestations of Raynaud's disease in other parts than the extremities are now looked upon with some doubt However, Raynaud himself, in his second publication and after mature consideration, described them as being a part of the disease He says "In the slight cases the ends of the fingers and toes become cold, cyanosed, and rigid, and at the same time more or less painful In grave cases the area affected by cyanosis extends upwards for several centimeters above the roots of the nails, at the same time the nose and ears may become the seat of analogous phenomena Finally, if this state is prolonged for a considerable time we see gangrenous points appear on the extremities, the gangrene is always dry, and may occupy the superficial layers of the skin from the extent of a pin's head up to the end of a finger, rarely more" One of his case histories describes changes in the nose and cheeks This case, Allen and Brown accept but Hunt rejects it because the trouble began a month after what may have been an attack of malaria However, the account is vivid and decidedly worth quoting

"Case VI Rose G, a washerwoman, aged 28 years, with fair skin and bright complexion, enjoyed habitually good health * * * In the month of March she had several attacks of tertian fever, which disappeared under a sedative Towards the middle of April she became very impressionable to cold

"Every time that she went out during weather at all cool, the nose, chin, cheeks, hands, and feet became pale, they

nal secretion. Some of the patients, if exposed, for instance, to the slightest embarrassment, would at once display cold purple fingers. Freeman, Smithwick, and White, in two combined researches, showed first that the denervated rabbit's ear was strikingly sensitive to the adrenal secretion and then went on to demonstrate that the arm of an individual deprived of all postganglionic sympathetic supply was equally sensitive. It only remained to find out why the same condition did not obtain, in the case of the leg, following lumbar sympathectomy. The explanation turned out to be anatomical. The customary removal of the second and third lumbar ganglia (the lowest ones receiving sympathetic *pre*ganglionic rami from the spinal cord) interrupts all *higher* sympathetic control of the blood vessels of the leg, but leaves the *post*ganglionic rami, emerging from still lower lumbar ganglia, intact. Thus the vessels of the legs are not left without postganglionic control and are not hypersensitive to the adrenal secretion. (See Figure 1.)

This explanation is necessary to account for the success of the procedures which were developed almost simultaneously by Telford in England and Smithwick in America. To free the arm from vascular spasm, all rami passing to the second and third thoracic ganglia are divided and the sympathetic cord is severed below the third ganglion. The rami of the stellate (inferior cervical and first thoracic) ganglion together with the second and third thoracic ganglia themselves are left intact (Figs. 7, 8, 9). Time seems already to have proved the correctness of these procedures which differ from each other only in detail and which free the arteries of the arm from any serious vasospasm, whether by exposure to cold or epinephrine injection. Moreover they offer the very decided advantage that if the stellate ganglion (first thoracic and inferior cervical) is left totally undisturbed, both as to its pre- and postganglionic rami, the unsightly falling in of the eye (enophthalmos) permanent narrowing of the lids and contraction of the pupil—*Horner's Syndrome*—will not occur. The avoidance of this disfiguring result, with its associated unpleasant sen-

cited by such a variety of stimuli that attempts to group them in a systematic way are as likely as not to increase rather than diminish the difficulty of diagnosis Spasm of the retinal vessels had better not be regarded as an exhibition of Raynaud's disease

Treatment—The background of treatment is the same as that of the mild and nonprogressive form of Raynaud's phenomenon, that is, avoidance of exposure to cold cold drafts and contacts, and, to preserve the natural warmth, the wearing of woolen clothing, mittens and stockings Ample shoes should be worn in cold weather The victim of Raynaud's disease will often begin the day by washing the hands in fairly hot water before taking the risk of entering an even cool room If she can be sure of avoiding exposure afterward and does not fear the habit, she may justifiably take a stiff drink of alcohol before breakfast! Whether physicians should advise such a course is a matter which the writer is not wise enough even to discuss

Sympathetic Ganglionectomy is now so universally if ungrammatically dubbed "Sympathectomy" that only a purist would avoid using the term Moreover, in the case of the arm, the approved procedure is no longer resection of the ganglionic chain but has gone back to what amounts to a combination of division of the chain and ramisection, as will presently be explained The story is this A very satisfactory and complete sympathetic denervation of the arm had been developed by Adson and Brown, following Kuntz's demonstration that the second thoracic ganglion often sent a sympathetic ramus to the lower end of the brachial plexus They removed, through a posterior approach, the inferior cervical and first and second thoracic ganglia But the arm, in many cases, only remained free from vascular spasm for a few weeks or months, though the completeness of the sympathectomy was vouched for by the total sudomotor and pilomotor paralysis It then appeared that the neurectomy was *too* complete, for the degeneration of all the postganglionic fibers left the smooth muscle of the arteries hypersensitive to the adre



FIGURE 7. UPPER THORACIC SYMPATHECTOMY—POSTERIOR ROUTE. Smithwick's Method. *A*, *B*, and *C*. Approach to the second and third left intercostal nerves *D*. Each nerve is cleared of all rami and local branches with a blunt hook and followed into the intervertebral foramen. *E*. Each nerve is gently teased out, exposing the sensory ganglion, and its roots are divided with scissors. The sympathetic chain is cut below the third ganglion and lifted up.

sations in the skin of the face and head, is decidedly worth while

Because the operations are now reasonably well standardized and are equally useful in the treatment of the various states of peripheral vasospasm which complicate other vascular diseases of the limbs, they are described below The indications in Raynaud's disease are that vasospasm should not be otherwise controllable and that the peripheral vessels, those of the digits in particular, should have been proved to relax sufficiently well in response to any effective test of reactive hyperemia (heating the body, paravertebral sympathetic block) That is, it must be possible to raise the skin temperature of the fingers (or toes) to, or nearly to, the high limit and that with reasonable promptness Otherwise the digital arteries are almost certain to be so fibrosed that the operation will do very little good

Sympathetic Ramisection, or Sympathectomy, for the Arm —Two routes are available the posterior approach, using a modification of the muscle splitting incision advocated by White, Smithwick, Allen, and Mixter, teasing out and dividing the second and third intercostal nerves, in order to eradicate completely their associated preganglionic (white) rami, and dividing the sympathetic cord below the third ganglion, as described by Smithwick, the anterior approach at the root of the neck, using the technique of Gask, dividing the sympathetic cord below the third ganglion, cutting the preganglionic rami to the second and third ganglia, turning up the upper stump and drawing it away from the spine to prevent regeneration, as advocated by Telford

The Posterior Approach, with Smithwick's modification, is here preferred

An incision is made about eight cm (three inches plus) long, and three cm from the mid line, nearly parallel to the vertebral column (sloping a little upward and outward) and centering upon the level of the second thoracic spinous process (third rib) The fibers of the trapezius muscle are transversely divided Those of the major rhomboid are split, as are those



FIGURE 7. UPPER THORACIC SYMPATHECTOMY—POSTERIOR ROUTE. Smithwick's Method. *A, B,* and *C* Approach to the second and third left intercostal nerves. *D.* Each nerve is cleared of all rami and local branches with a blunt hook and followed into the intervertebral foramen. *E.* Each nerve is gently teased out, exposing the sensory ganglion, and its roots are divided with scissors. The sympathetic chain is cut below the third ganglion and lifted up.

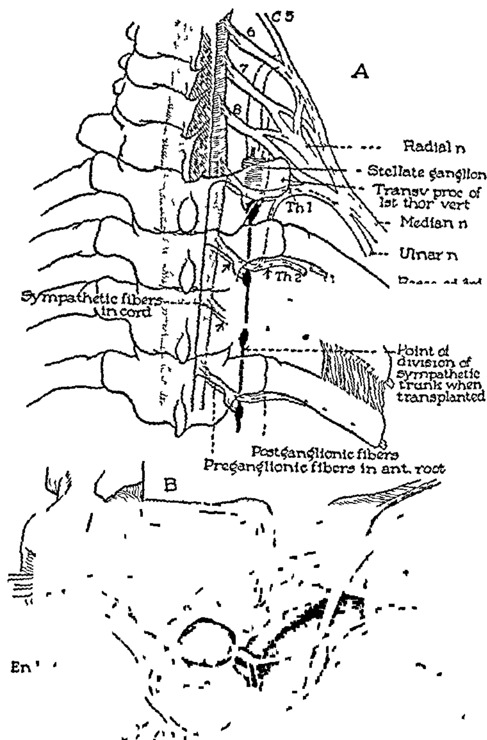


FIGURE 8 UPPER THORACIC SYMPATHECTOMY—POSTERIOR ROUTE *A* Scheme to show plan of preganglionic sympathetic interruption (indicated by arrows) Preganglionic rami and sympathetic chain in black No pre or post ganglionic rami of the stellate ganglion are disturbed *B* Cross section at level of third left intercostal nerve Operative approach

of the sacrospinalis muscle. The third rib is exposed, isolated, and resected, its proximal end and the corresponding spinal transverse process being subsequently bitten off with the rongeur.

The pleura is pushed away from the vertebral column with gauze.

A bright, brain-spoon retractor serves to reflect light toward the vertebral bodies, or an illuminated retractor is used.

The third intercostal nerve is isolated by blunt dissection, divided at a convenient distance from the vertebrae, followed in to its exit from the vertebral column and freed from all attachments with a dental spatula. In so doing, the posterior root ganglion, with the anterior and posterior roots, are teased out, brought into view and divided. In this way all preganglionic fibers are thoroughly interrupted. A leak of cerebrospinal fluid is unusual.

The second intercostal nerve is similarly treated. In the meanwhile, the sympathetic ganglionated chain has not been disturbed.

The sympathetic is now picked up with a blunt hook between the second and third thoracic ganglia as it lies against the vertebral column and is divided with scissors below the third ganglion. Its upper portion is now turned up, care being taken not to disturb the stellate (first thoracic and inferior cervical ganglia) ganglion. The object of this step is to obviate Horner's syndrome (contracted pupil and enophthalmos) and leave all outgoing postganglionic rami intact. The stump is attached by a silk stitch to any convenient fascial structure. The wound is closed in layers with silk. If the pleura is injured, the opening is covered with a fragment of muscle pressed down with gauze.

The Anterior Approach (Gask's with Telford's modification) is made through an eight cm. (three inch plus) incision, two fingers' breadths above and parallel to the clavicle. A cervical and brachial plexus block plus local infiltration with procaine, rather than a general anesthetic, can be used if desired.

After cutting skin and platysma, the clavicular portion of the exposed sternomastoid muscle is divided and also the little mylohyoid This step gives access to the anterior surface of the scalenus anticus muscle upon which the phrenic nerve lies The muscle is cleaned, the phrenic is retracted mesially with a tape and the transverse cervical vessels severed if necessary After the scalenus anticus has been isolated and divided two cm above its insertion into the first rib, a view is had of the subclavian artery and the brachial plexus The latter is severely let alone, not cleared at all The carotid sheath is retracted mesially

Before the subclavian artery can be pressed downward, the thyro cervical axis, which originates from its convexity, must usually be divided between good sized silk ligatures On depressing the subclavian, Sibson's pleurocostal ligament, which holds the pleural apex against the lower border of the first rib, is revealed When this has been cut, the pleura falls back and is further peeled away from the costovertebral angle by gentle pressure with gauze

The wound is now deep and must be lighted by an illuminating retractor or a reflecting spoon The stellate ganglion is seen just behind the origin of the vertebral artery, between this vessel and the head of the first rib The chain, below it, is carefully exposed by detaching any loose tissue from its surface The stellate ganglion must not be disturbed When the chain has been cut, with long curved scissors, below the third ganglion, the upper stump is lifted upward sufficiently to divide the rami entering the second and third ganglia It is then attached to any convenient structure with a fine silk stitch The retractors are now removed and the pleura allowed to fall back

The only parts requiring suture are the clavicular portion of the sternomastoid muscle—to restore the contour of the neck—and the platysma The skin is approximated with clips

Both sides can be operated upon at one sitting provided the operator is sufficiently familiar with the procedure The patient usually is up and about on the third to fifth day

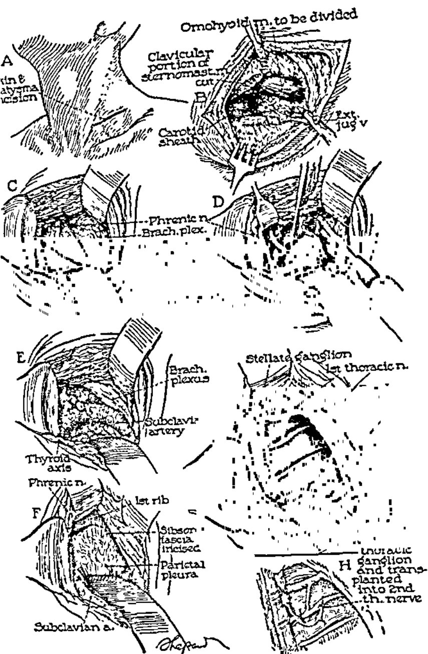


FIGURE 9. UPPER THORACIC SYMPATHECTOMY—ANTERIOR ROUTE. Method of Gask and Telford. *A, B, C, D,* and *E,* approach. After *E,* it may be necessary to divide the thyroid axis, in order to depress the subclavian artery. *F.* Division of pleuro-costal ligament and depression of pleura. *G* and *H.* All rami of second and third ganglia are divided, sympathetic is cut below the third and planted into second thoracic nerve (De Takats).

Lumbar Sympathetic Ganglionectomy —Though this procedure can be carried out through either a transperitoneal (abdominal) or retroperitoneal approach, the former is never considered at present unless a bilateral operation is contemplated and there is some other reason for opening the abdomen The transperitoneal procedure is not only much more disturbing to the patient than even a bilateral extraperitoneal attack but the sympathectomy, especially on the right, is more difficult

The incision is very much that of an approach to the kidney Flothow recommends a muscle splitting one, dividing the skin almost horizontally just below the twelfth rib, then passing forward and downward, splitting the oblique muscles in the direction of their fibers (and transsecting at least a few of the fascicles of the internal oblique) The transversalis and its fascia again are split horizontally, care being taken not to open the peritoneum The operator then pushes retroperitoneally toward the vertebral column, lifting forward the lower pole of the kidney and the ureter (which he does not see)

The first landmark upon the surface of the psoas muscle, before the sympathetic is approached, is the genitofemoral nerve, a straight, tense, white filament Beyond this, and close against the anterior part of the exposed vertebral bodies, lies the faintly pinkish gangliionated chain solidly attached by its fine rami

On the right, the vena cava must actually be retracted to expose the chain On both sides, the renal fascia is seen at the upper angle of the dissection The renal artery can if necessary be felt

Two rather spindle shaped but not strikingly marked ganglia will usually be found, the second and third lumbar, the latter just above the common iliac vein (on the right) or artery (on the left) Occasionally the two are fused The ganglia and chain vary in size and their exposure may be made difficult by the extent to which the great muscles overlap them as they are applied to the vertebral bodies The peritoneum must be held forward by gauze packing and a broad curved (lighted)



FIGURE 10. LUMBAR SYMPATHECTOMY—RETROPERITONEAL ROUTE. In B, some tendinous fibers of the internal oblique (not shown) will have to be divided. In E, the gangliated chain is too distant to show that the rami approach L, 1 from above, L, 2 horizontally and L, 3 from below. L, 2 and 3 are often fused

retractor It is best to sever the rami first, then cut the sympathetic with scissors below the third ganglion It can now be lifted up and divided above the second just below the renal vessels To secure a sympathetic denervation which will carry the vasomotor paralysis as high as the thigh, the first lumbar ganglion must be included in the resection In women, this is harmless in men, it is very likely to upset the mechanism of

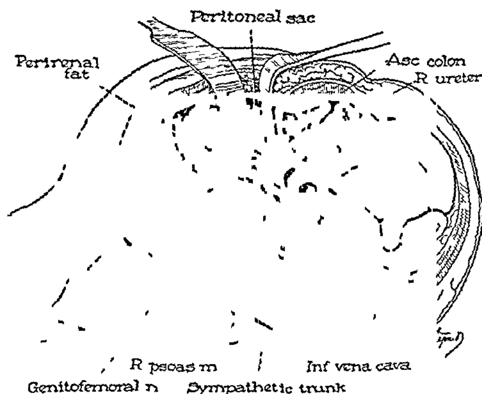


FIGURE 11 LUMBAR SYMPATHECTOMY A cross section showing the retroperitoneal approach to the right sympathetic chain The displacement of viscera, including the vena cava, is not exaggerated The psoas muscle must be depressed to expose the sympathetic

ejaculation Removal of this ganglion in males should therefore be practiced with caution

The wound is closed with silk in layers

The result of sympathectomy should ideally be the establishment of warmth, dryness and freedom from vasospasm in the affected limb Tests of the skin temperatures should show a degree of vasodilatation which can neither be increased by

any attempted reactive hyperemia nor appreciably diminished by moderate exposure to cold. In other words, the power of the peripheral circulation to display a vasomotor reaction in either direction should be lost. The condition should be permanent. In the legs, it usually is permanent. In the arms, under the operative system now in use, it will probably prove so to be. Strangely enough, persons who have lost their vasomotor control do not seem to miss it. They enjoy the sense of warmth in their fingers or toes and make no objection to the dry skin. Even if the procedure is not completely successful, patients are usually pleased with the result; and the individuals actually made worse by degeneration of the postganglionic vasoconstrictive nerves and exposure to spasm from adrenal influence (under the earlier method of treating the supply to the arm) are after all few.

Pneumatic Hammer Disease.—This form of arteriospasm, though much like Raynaud's phenomenon, affects only male workers who use a rapidly vibrating stonecutter's hammer. Within a few months after first using the instrument, the workman may become subject to attacks of vasospasm in certain fingers. These attacks are excited by exposure to cold, particularly in the early morning or after the day's work is over—never while the hammer is actually being used. Between attacks the fingers show no change, nor is the disease progressive or destructive. Because of the manner in which the hammer is held, the fifth, fourth, and, occasionally, the third finger of the left hand are especially affected; sometimes the tips of those of the right hand as well. The disease was first identified (1917–18) by investigators for the United States Department of Labor and of Public Health. A good description is given by Hardgrove and Barker, from the Mayo Clinic.

Scleroderma Acrosclerosis Sclerodactyly

This disease, actually rarer than that of Raynaud, may possibly be a near relation. It is practically confined to young females, showing itself in childhood. Attention has been called

to the hardening of the subcutaneous tissue, shrinking of the skin and smoothing of the natural wrinkles in advanced Raynaud's disease Such a state is known as Sclerodactyly Scleroderma seems, in some instances, to be a more wide spread development of the local digital vasospasm and secondary sclerosis, in others, the general hardening of the subcutaneous tissues and atrophy of the skin appear to be primary A form is actually described in which the skin of the body and limbs becomes hairless, thick and firm, the process spreading down the arms to the hands without causing vasomotor symptoms In any case, the full fledged disease stiffens the fingers, forearms, and even upper arms, makes a mask of the face, smoothing out all natural folds and wrinkles, tightens and fixes the skin, particularly over the forehead and malar prominence, and finally, in many cases, leads to arthritic changes and ankylosis The eyes and mouth are reduced in size Neither can fully be opened, nor can the eyelids be tightly closed The front of the chest is apt to be affected, the toes often, the feet only occasionally

Microscopic examination reveals atrophy of the growing layer of the epidermis The deep skin is fibrosed, the subcutaneous tissue as well Indeed, fibrosis of soft tissue is sometimes so advanced as to extend into the muscles and bind skin to bone The small arteries are embedded in scar, their caliber greatly reduced This extensive hardening of the tissues may follow upon Raynaud like attacks of cyanosis of the fingers—in which case it follows rather promptly—or it may occur coincidentally with such attacks It may even fully establish itself in the absence of any attacks whatever It is idle to speculate whether arterial spasm is a prime factor or whether some basic infection or endocrine disturbance is at the bottom of the whole process

The course of the disease, though often rather rapid in its early stages, during which the hands, arms and face are hardened, is decidedly chronic, dragging along for years toward a fatal ending Once hide bound and stiffened by arthritis, the victim is in a pitiable and hopeless state No one actually

recovers from the disease. The best that can be asked is that it should become stationary at a stage when the individual is not actually crippled.

Treatment is most unsatisfactory. When intermittent vasospasm is a feature, the patient can of course be protected from cold and other vasospastic influences, as in Raynaud's disease. The skin is softened and made as pliable as possible by massage and greasing. Nutrition is kept up as well as possible. Sympathectomy occasionally makes the fingers more supple and comfortable, though any decrease of stiffness is rather more likely to be due to diminished edema than to any increased pliability of the skin. It should not be used unless vasodilatation has been proved possible. If the metabolism is low, desiccated thyroid should be pushed.

Arterial Spasm in Response to Injury

Traumatic arterial spasm is brought on by a variety of injuries, many of them violent; such, for instance, as wounds of war and accident, bullet wounds, fractures, blows, stabs, and punctures. Sometimes there is evidence of direct trauma to the arterial wall; almost as often, surprisingly enough, the artery itself seems never to have been touched. Large arteries such as the femoral or the brachial have chiefly been affected, partly perhaps because of their length and exposed position. On the whole, it would seem as if the spasm were the result of a disturbance of the local vasomotor nerves, and at the beginning, at least, thrombosis is certainly not a factor. Spasm of this sort lasts for hours or even days and usually leads to no complicated after-effects. There is another great group of arterial vasospasms, however, undoubtedly merging into this one, which is of a decidedly more chronic sort. In this, the spasm appears as a rule to be due to a vicious reflex, and the associated changes in the skin, nerves, muscles, bones and joints may overshadow the direct evidence of vasoconstriction. Included in this group of chronic arterial spasms are causalgia and the causalgia-like states, reflex dystrophy of the extremities and traumatic osteoporosis. There are still other

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pin prick. Motions of the toes were feeble and painful. It was thought that all this was due to contusion by the bullet, causing an intense spasm of the femoral artery. Accordingly, the leg was massaged every fifteen minutes, and after nine hours the whole extremity became bright red and warm, the pulse returned and the normal sensibility was restored. The wound healed without incident.

Here, of course, are instances of contusion of an artery by a bullet which passed very close to it; in effect, a blow upon the vessel itself yet without any permanent injury or thrombosis. Other reports such as that of Kuttner and Baruch tell of examinations of such vessels and in one instance (a wound at *the ankle*) excision of a seemingly thrombosed stretch of posterior tibial artery. Whereupon the vessel, both proximal to and *beyond* the part resected, resumed its pulsation (retrograde circulation) and much to the operator's surprise, no thrombosis in the excised specimen was found. Thus *additional* information was secured; for it is evident *that resection* of the contracted vessel broke up the spasm of the vascular tree of which the posterior tibial was the *main stem*.

Instances of acute arterial spasm resulting from fractures are described by Montgomery and Ireland, from whose paper the following is abstracted:

A boy, four years old, had suffered a fall upon his left elbow an hour before coming under observation. There was found a fusiform, discolored swelling with lateral angulation of the arm below the site of injury and marked tenderness over both condyles of the humerus. Distal to the elbow, the skin was *dead white* and distinctly colder than that of the opposite hand and forearm. No pulsation could be palpated in the left radial or ulnar arteries. The X ray showed a comminuted supracondylar fracture with lateral displacement of the lower fragments, the radius and ulna being dislocated posteriorly.

Four and one-half hours after the injury, reduction under gas oxygen was carried out. The circulation remained unchanged.

states such as acute arteritis, seemingly related to an anatomic abnormality such as cervical rib, which are even less easily classifiable All these are roughly sorted out below in the sections which follow It is not necessary that all should agree as to their relation to each other

Acute Traumatic Arterial Spasm Arterial Stupor—Though it would be impossible to tell of all the injuries which may excite traumatic spasm, several cases, due to bullet wounds, to fractures, to punctures and to blows have been selected as examples It will be observed that the first cases cited are of *a rather direct type of trauma to great vessels, and* are marked by arterial stupor Naturally, most observations of this sort have been made in war time Two very instructive experiences are reported by Kroh (1915) The first is one of a wound of the right inguinal region by a revolver bullet There had been a good deal of venous bleeding from the wound of exit The femoral artery was pulseless The soldier complained of poorly localized pain in the lower thigh Exploration showed that the saphenous vein was severed This was doubly ligated The femoral arteriovenous sheath was seen to be infiltrated with blood The femoral vein, fairly *well filled*, was identified but the artery could not be found for some time Finally a pulsating thread, the size of a knitting needle, was isolated which *should have been the femoral but seemed* to be an anomaly! As it was watched, however, it gradually enlarged and much to the operator's surprise soon took on *the size and pulsations of the femoral*

This observation evidently prepared Kroh for the following case which was treated without exploration A soldier, who had been shot with a rifle bullet through the upper thigh, complained of numbness and tingling in his foot The bullet entered near the anterior superior spine and came out just below the scrotum The intervening skin was discolored with blood The pulse in the femoral artery just below the track of the bullet was barely perceptible In the popliteal and posterior tibial it was absent The skin from the knee down was yellowish white, *cold and damp, completely anesthetic*, even to

Hence the multiplicity of names. Whether the basic exciting lesion is in the sympathetic or sensory nerves and just what reflex pathways are involved is still uncertain. Doubtless many sorts of injury, including blows, infected and uninfected wounds, even burns and frostbites, are able to set off the prolonged, serious, but usually reversible reflex disturbance. Although he invokes, even in one case, both a vasodilating as well as a vasoconstricting influence, Leriche has consistently laid the trouble to the vasomotor nerves, the peripheral sympathetic system. A typical case of his, of which another example is illustrated opposite page 148, is here quoted:

"Case 3. This man is a polish worker. After a severe blow on the left foot he presented inability and pain. There was no fracture. No treatment gave any relief. The patient stayed in bed, and for almost a year could not walk. He was finally considered to be a malingerer and sent to me with this diagnosis.

"Examination showed the whole left lower limb to be extremely painful: the slightest touch upon the skin made him cry out. No movement was possible. The leg and the foot were cyanotic. There was evident atrophy of the foot, the leg, the thigh [illegible] of the leg.

"Considering this wide disturbance of his troubles, I performed a lumbar ramisection on Nov. 22, 1926. The same evening pain and cyanosis had gone. The following day feeble movements were possible. Sixteen days after operation, the patient got up for the first time for eight months. He could bear weight upon the foot without suffering. Functional conditions improved progressively. The bone recovered rapidly its calcareous matter, and successive roentgenograms showed a very regular reconstruction of the bone which was absolutely normal after three months. At the end of March, 1927, the patient was discharged perfectly cured."

In other instances of this general type, Leriche makes a great point of the traumatic arthritis and hydrops of joints

Incision was then made into the antecubital region where a large hematoma was found and evacuated The brachial artery was seen to pulsate down to a point an inch proximal to its bifurcation, but here all pulsations abruptly ceased "as though the blood were *striking against a solid structure*" The pulseless arteries were contracted, did not appear to have been injured in any way and seemed not to be thrombosed. Warm, wet dressings were applied to the open wound

On the day following, the hand felt warm and the nails were no longer completely blanched, but the pulses were still absent Three days after operation, a faint radial pulse returned, *slowly improving in quality* until it became altogether equal to that of the uninjured arm Eleven days after operation, the wound was closed and a posterior cast applied However, no attempt at final reduction was made at this time Indeed it was not until fifty three days after the injury that complete replacement by an open operation was made This caused no circulatory upset

It is not easy to say whether this spasm (stupor) was induced by direct trauma to the artery, of which the hematoma perhaps offers some evidence, or whether injury to its sympathetic nerve supply was the cause Perhaps it makes very little difference, since, in any case, a local nervous mechanism in near relation to the vessel would seem to have been involved *However, such an arteriospasm is a direct result* of the bullet wound, fracture, or whatever, and not a reflex disorder, as seems to be the case with the more obscure and complicated group which follows

Reflex Arterial Spasm Chronic Segmental Arterial Spasm Causalgia Reflex Dystrophy of the Extremities Trophic Edema Traumatic Osteoporosis—Although this group is a loose one and although observers, dating back to S Weir Mitchell, may perhaps have been describing a considerable variety of states, there has been for some years a fair unanimity of opinion that there is such an entity as prolonged reflex arterial spasm and that this is capable of causing disorders of skin, connective tissues, nerves, muscles, joints and bones

Hence the multiplicity of names. Whether the basic exciting lesion is in the sympathetic or sensory nerves and just what reflex pathways are involved is still uncertain. Doubtless many sorts of injury, including blows, infected and uninfected wounds, even burns and frostbites, are able to set off the prolonged, serious, but usually reversible reflex disturbance. Although he invokes, even in one case, both a vasodilating as well as a vasoconstricting influence, Leriche has consistently laid the trouble to the vasomotor nerves, the peripheral sympathetic system. A typical case of his, of which another example is illustrated opposite page 148, is here quoted:

"Case 3. This man is a polish worker. After a severe blow on the left foot he presented inability and pain. There was no fracture. No treatment gave any relief. The patient stayed in bed, and for almost a year could not walk. He was finally considered to be a malingerer and sent to me with this diagnosis.

"Examination showed the whole left lower limb to be extremely painful: the slightest touch upon the skin made him cry out. No movement was possible. The leg and the foot were cyanotic. There was evident atrophy of the foot, the leg, the thigh, the buttock, and even the left side of the back. Radiography showed extensive decalcification of the foot and lower third of the leg.

"Considering this wide disturbance of his troubles, I performed a lumbar ramisection on Nov. 22, 1926. The same evening pain and cyanosis had gone. The following day feeble movements were possible. Sixteen days after operation, the patient got up for the first time for eight months. He could bear weight upon the foot without suffering. Functional conditions improved progressively. The bone recovered rapidly its calcareous matter, and successive roentgenograms showed a very regular reconstruction of the bone which was absolutely normal after three months. At the end of March, 1927, the patient was discharged perfectly cured."

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resulting from what he believes to be reflex *vasodilatation* Such leave behind many stiff joints Whether or not he confuses the passive congestion of vasoconstriction with vasodilatation is immaterial The disorder of joints, bone and muscle (atrophy) presumably go together as a rare response to even rather mild injuries

Another somewhat similar type of disease but now due to an infected wound is described by Lehman D E D, a school girl of eighteen, had suffered, four months before coming under observation, a punctured wound by a crabapple thorn upon the ulnar border of the right forearm This had become infected and had been subjected to exploration and drainage A sinus had been left surrounded by induration, heat, and redness half way to both wrist and elbow The patient feared to have the forearm touched and could barely move the wrist and fingers During the following seven months, four operations for drainage were performed Almost a year after the original injury, the forearm and upper arm were swollen and doughy up to the axilla Pain was continuous The fingers were cool The right radial and brachial arteries pulsated (to the touch) less strongly than those of the left hand Though one of the old incisions was unhealed, the temperature and leucocyte count were normal Amputation had been suggested

On the basis that the condition resembled the "trophic edema" of some war wounds and because tissue removed at the last operation had shown a perivascular inflammation, a periarterial sympathectomy was performed upon the right brachial artery Two days after the operation, movements of the fingers had improved and tenderness of the forearm had diminished Six weeks later, motion at the elbow was free and that of the fingers was beginning to improve Swelling had disappeared and the skin had become warm and dry The wound had healed In six months, except for some weakness, the extremity was normal

This case appears to have been one of a rather diffuse vasospasm in response to a local injury and infection which probably did not actually touch any large artery or nerve The

vasospasm mounted well above the lesion and affected both motor and sensory functions. The reflex disorder, for such it must be called, was broken up by removing the nerve fibers surrounding the brachial artery. It would seem that a cure was due to the interruption of stimuli going *toward* the central nervous system, since the interruption of sympathetic impulses passing out upon the brachial artery could hardly have affected parts of the arm proximal to the operative field. This case, in respect to its painful state and oversensitive skin, much resembles causalgia, an instance of which is taken from the original publication of Mitchell, Morehouse, and Keen (page 107).

"Case 24.—Hiram Weston, aet. 42, Co. E, 18th Mass., enlisted May, 1861. Healthy until wounded, in the Wilderness, May 5, 1864. He was running at a double quick, and was shot in the left arm. The ball entered three and a half inches immediately above the internal condyle of the humerus. It emerged directly below the anterior angle of the axilla, two and a half inches lower. The ball passed over the nerves, and injured the ulna nerve especially.

"He felt violent pain throughout the limb, which was instantly flexed at every joint and so continued for fifteen minutes, when it was extended by the aid of the other hand. The pain which then began never left him. The arm soon lost motion entirely: but within a few days regained so much as it has now. As to sensation, he can tell us nothing, except that probably it was only damaged in the ulnar range of nerve supply.

"*Present state.*—It is now fifty days since this man was shot. Of the intervening period, he gives a very clear account. Immediately after the wounding, the whole limb swelled; but this rapidly subsided and the hand was no larger than its fellow, until about the fortieth day, when it became rapidly oedematous. The pain has consisted all along of darting pangs from below or under the elbow, down into the hand, but not on the anterior surface of the forearm. In the hand, the pain is burning and tingling, or as he calls it 'prinkling'. • • • The

hand is sore to touch everywhere, but tact is unimpaired, save in the little finger and ulnar side of the ring finger * * * The nutritive changes did not become marked until about the forty fifth day, they are now obvious, and in time will doubtless give rise to the glossy skin, to which we have so often referred The hand is swollen The palm is red and dotted with patches of thickened epithelium. * * * The nails are laterally much arched, the skin at their bases retracted, * * * The joints are exquisitely tender, and very stiff and swollen The patient has kept the hand wet ever since he was hurt * * * Tactile sensation is perfect throughout the hand, except in the fourth finger and the ulnar side of the third "

The authors feel the case to be typical of the symptom complex which Mitchell later named causalgia. The burning pain comes first, then the glossy skin But the pain itself only arises during the healing of the wound Moreover it may transfer from the field of the wounded nerve to that of an unwounded one Its site is always the foot or hand and here the nutritive changes are seen The part affected is not only subject to the intense burning but is so sensitive to a touch or draft of air that the victims almost invariably keep it moist.

The foregoing description, which does little justice to the full account of Mitchell and his associates, fits fairly well the chronic arterial spasm and reflex dystrophy of today Into the story of Hiram Weston one may read, if one likes, an ulnar nerve injury, a blow upon the arteriovenous bundle followed first by a venous thrombosis and, after some forty days, by the full fledged edema, malnutrition, and oversensitiveness of a serious, extensive, reflex arterial spasm. One can find in the writings of Meige and Madame Athanassio Benisty descriptions of causalgias resulting from wounds received in the war of 1914–18, exactly similar to those of Mitchell But the French neurologists call attention to the fact that Weir Mitchell described particularly wounds of the brachial plexus and failed to notice the causalgias of median and sciatic nerve injuries They themselves describe these states very accurately They say

"In the painful form of wounds of the median nerve with major causalgia, the hand takes on an emaciated aspect; the skin is delicate, wrinkled, rosy in color, and marbled with bright red patches; it is hot both subjectively and objectively. The arterial pressure is elevated. Sweating is abundant. Nutrition is decidedly changed."

They find that the nerve itself, when exposed, is congested and that the vasomotor and sudomotor disorder is shown principally in the structures supplied by nervous terminals of the sympathetic; that is, in the various nervous (Pacinian) corpuscles and the capillaries of the skin. They regard the median and sciatic as especially vulnerable because they are rich in sympathetic fibers and are well supplied with vessels which themselves are abundantly furnished with vasomotor nerves. There is probably a distinction, which has never been clinically clear, between the causalgia of median or sciatic origin, that is, a primarily nervous lesion on the one hand, and on the other, the sort of injury which Leriche was the first to cure by interruption of the periarterial nervous pathways. Both his early cases were wounds in the region of the upper axillary artery. In one, he stripped the axillary and later resected, with a very favorable result, its proximal portion. His second case is briefly described below:

Corporal G. was wounded Sept. twenty-fifth, 1915, by a bullet which fractured his left clavicle. A flaccid paralysis of the left arm made it seem probable that the brachial plexus had been divided. Ten days later, Leriche explored the plexus, finding it and the distal part of the subclavian artery embedded in scar tissue. The plexus had not actually been injured. There were no pulsations below the clavicle. He dissected free the plexus and vessels without effect. The hand remained cold, deeply cyanotic, and totally paralyzed.

Five months later, the soldier reported back to Leriche. The hand was colder and more purple than *ever*. The skin was glossy and felt cold like that of a snake (by contrast with some of the median and sciatic nerve injuries in which the extremity is hot). There were blood-blisters on the *fourth*

and fifth finger-tips Crises of burning pain kept recurring in the hand Leriche now laid bare the upper brachial artery which was only two to three mm in thickness, did not pulsate, and was covered with reddish patches The neighboring nerves were soft Stripping off the outer coat of the artery caused no bleeding whatever But on the following day the soldier noticed tingling in his whole arm and felt much relieved The left hand, previously colder than the right, had now become the warmer of the two The blisters dried up Three weeks later some motion was beginning to return in the forearm No further account is given

Leriche believes that the wide-spread arterial spasm broken up by periarterial sympathectomy accounts for the paralysis and atrophy in such cases, no actual lesion of the great nerves being present Yet the vasomotor disorder seemingly is capable of affecting the peripheral nerves through their blood supply, causing paralyses and weird contractures The fingers, for instance, may be left extended but pointing together, the thumb flat in the palm, the wrist straight or flexed Such a deformity somewhat resembles that of paralysis of the median nerve above the elbow With it, the trophic disorders so often seen—blisters, ulcerations, desquamation, deformed nails—are consistent Probably some causalgias or causalgia like states are primarily due to peculiar peripheral nerve injuries, while others are the result of irritations of the plexus of nerves surrounding the great arteries of the limbs In the latter case, vasomotor spasm, if sufficiently prolonged, may cause a great variety of nutritive changes in bones, joints, muscles, subcutaneous tissues and skin, imitating in some cases, if it does not actually occasion, serious inflammation of certain great nerves Apparently a vicious sensory sympathetic reflex is set up, as pictured by De Takats Indeed, some such mechanism must be imagined to explain the dramatic cures so often secured by blocking the periarterial nervous pathway or excising the sympathetic rami and ganglia

Minor degrees of these reflex disturbances are rather common It is not necessary that all or indeed many of the peculiar

changes described should be present. A little edema, a little alteration of superficial sensibility so that scratching or handling the part is unpleasant, a diminution of the peripheral pulse, a moderate atrophy of the bones; such will often be noticed after a variety of disorders ranging from serious fractures to cat-bites and from laceration of a great nerve to a superficial bruise. The condition may show a tendency to spontaneous recovery, but as a rule it is decidedly persistent. The best test of its presence is a paravertebral nerve block (or spinal anesthesia). This will usually, for the moment, bring on a full vasodilatation, restore the natural sensibility of the skin, and even diminish edema. It will guarantee a favorable result from sympathectomy, provided the seriousness of the symptoms demands such a step.

Treatment.—In all reflex osteoporoses, reflex edemas and causalgia-like states, the effect of paravertebral nerve block with procaine should be studied. In the milder cases, hypersensitiveness of the skin will not only disappear for as long as the block lasts, but will sometimes, from that moment, show a progressive improvement. It may therefore be advisable to repeat it. Relief is favored by the skillful use of massage, heat and even [illegible] curative. But in the more serious cases, a sympathetic neurectomy is required. Obviously a paravertebral ramisection or ganglionectomy will break the outflow of vasoconstricting impulses, and there are very few cases incurable by this means. However, as already explained, even a "periarterial sympathectomy", that is, stripping off the outer coats of the principal artery supplying the affected limb, which presumably acts by interrupting mainly central-going impulses, will often break up the vicious reflex. This was Leriche's original contention and is well demonstrated by Lehman's case. The whole matter will come up again in the management of the remarkable arterial spasms which are so often associated with arterial embolism and even with venous thrombosis.

Acute Arteritis: Cervical Rib: Scalenus Anticus Syndrome

(Naffziger Ochsner).—There is some question whether all these terms refer to the same abnormal sort of arterial spasm. Acute arteritis may perhaps be a variety of arterial stupor or even reflex chronic arterial spasm It will be sufficient to present it as occurring in the brachial artery Although infection at a distance has been invoked as a cause, it is held here that some unnatural relation of the brachial plexus and subclavian artery to the first rib or an actual cervical rib is more likely to be responsible

The Clinical Signs of abnormal pressure upon or irritation of the brachial plexus and subclavian artery usually point to *a nervous rather than to an arterial difficulty Indeed, Naffziger* regards the arterial disorder as relatively rare The syndrome is more common in women than men

The principal nervous symptom is pain, referred to the supraclavicular region and shoulder, the side of the neck and ear, the arm and forearm, especially upon the ulnar surface It may be tingling and numb or sharp and knife like It is usually aggravated by depression of the shoulder Supraclavicular tenderness and a radiation of the pain down the ulnar side of the arm are often brought on by pressure over the scalenus anticus muscle at the root of the neck

The first vascular manifestation is usually weakness of the arm, made worse by exercise Then, coldness, numbness and tingling gradually set in An early diagnosis is difficult Relief of the distress by elevating the shoulder and aggravation of the symptoms by lowering it are confirmatory The radial pulse may be obliterated or its weakness may be evident, or oscillometry alone will show that pulsation is slightly lessened in the affected arm The brachial artery is occasionally felt as a tender cord, proximal to which the axillary and subclavian beat normally If necessary, arteriography can be used

In a case described by Clute the following observations were made The patient, E K, a man, thirty five years of age, had suffered for several years from a nonspecific prostatitis Six months before coming under observation, he had noticed blanching, coldness, and occasionally cyanosis in the left hand

and arm. Pain was moderately severe and had progressed upward along the radial and brachial arteries. The left arm was found to be cold, damp, and cyanotic. On motion, the fingers blanched. The brachial artery could be palpated as a deep, tender, swollen cord. There was a good pulse in the axillary, none below.

Clute first explored the supraclavicular region. The subclavian artery and brachial plexus did not appear to be pinched between the scalenus anticus muscle and the first rib. Nevertheless, a few of the fibers of the tendon were divided. No improvement followed.

The brachial artery was then explored and found to be a firm cord surrounded by a mild edematous reaction. Two inches of the artery and vein were resected. The vein proved to be normal; the artery, the seat of "chronic periarteritis". There was no thrombosis and no bacteria could be cultivated from the tissues. Improvement set in within three days and ended in almost full recovery of all functions. The arm was only slightly disabled but the radial pulse did not return.

Here is a disease which is not associated with injury, with which infection can have had little to do, and which may fall into the class described by Telford and Stopford (who attribute the explanation of the condition to Todd) namely, that either a first or a cervical rib is capable of making pressure upon and irritating sympathetic fibers entering the lower cord of the brachial plexus for distribution to the brachial artery (rather than the subclavian or axillary). The initial vasoconstriction causes pallor and coldness. Later, they say, the vasa vasorum are obliterated and finally the brachial becomes thrombosed. They feel that it is not so much the abnormal position of the rib as a peculiar exposure of certain sympathetic fibers to injury which causes the trouble. The periarterial filaments are in the state of irritation which Leriche has so vividly described by saying that the artery has become "a diseased sympathetic nerve".

It is perhaps unfair to suggest that the particular case just described is really one of an unnatural relation between the

upper rib and the brachial plexus. Yet the signs and symptoms are those characteristic of the circulatory disorder caused by a cervical rib or the scalenus syndrome of Naffziger. The truth is that only after many years and because of repeated lowering of the shoulder or bending of the head, does the necessary nervous irritation due to contact between rib and the lowest part of the plexus take place. A cervical rib may be present on one side and the symptoms on the other! Ochsner finds that in many cases the scalenus anticus appears to have become unnaturally shortened (by repeated irritation of the nerve supplying it) and to have lifted the cervical or first rib unnaturally high. Thus a vicious circle is established which can be broken by dividing the tendon of the scalenus anticus and letting the rib drop. Adson and Coffey had already suggested this step but with the idea of letting the artery and plexus slide forward and downward. This matter will again come up in a consideration of treatment. When exposed, at operation, the subclavian artery, *distal to the rib*, has often been noted to be dilated, almost like an aneurysm. The dilatation is presumed to be related to the constriction of the brachial beyond, which, by the time the signs of the disease are advanced, may have become impermeable because of contraction or actually thrombosed.

Treatment.—When the signs persist, even though the use of the arm in a hanging posture is prevented and in spite of sleeping with the arm suspended above the head, operation is indicated. The supraclavicular region is approached as in the anterior operation for cervico-thoracic ramisection (page 127) and the subclavian artery can readily be inspected, but it is not easy to be sure that the supposed nervous irritation is taking place unless a cervical rib is present, the first rib is held very high, or the subclavian artery is enlarged. In any case, the phrenic nerve can be drawn aside and the tendon of the scalenus anticus divided. A lowering of the rib may be the only obvious change secured, in which event the wound may properly be closed without further surgery, to determine the effect of the procedure. In an early case, the brachial artery

will, at once or in a few days, relax. If it remains a contracted cord, it had better, as Leriche directs, be resected for several inches. Whether or not the radial pulse returns, the symptoms are then likely to be relieved.

Permanent States of Vasospasm

Up to this point, the arteriospasms described have been intermittent, temporary or prolonged, yet as a rule not strictly permanent. Those now to be considered are of a permanent sort. Some are due to such disabling diseases of the central nervous system as infantile paralysis. Others are very similar permanent states of vasospasm for which no cause can be named. Why some of these vasoconstrictions should result in ulcerations, some in trophic disorders, and others merely in blue, sweating feet and hands, by contrast with the vicious causalgias just described, is as yet not understandable. One can only keep in mind the simplest form which arterial spasm may take, and recognize the variations upon it.

Infantile Paralysis: Spastic Paralysis.—Children born with [illegible] cold, damp limbs. That this unpleasant state is due to arterial vasospasm is proved by the vasodilatation which re- [illegible] the vasoconstriction is due primarily to disuse or to a neurogenic disorder is not clear. In any case, it [illegible] This was [illegible] a rami- [illegible] to diminish the tonicity of the muscles in spastic paralysis. The treatment failed in its principal object but was discovered quite unexpectedly to have left the paralyzed limb warm, pink and dry. Actually it opened the way for the general use of sympathetic ganglionectomy to relieve arterial spasm. Not only is the blue, cold extremity made pink and comfortable but if the atrophy of bone and soft parts has not

existed for too many years, the limb, whose length and girth have lagged far behind its mate, may show an acceleration of growth The procedure is also of use in connection with plastic operations on the foot, encouraging the healing of wounds To determine that the operation is indicated it is only necessary to use one of the tests of vasoconstriction described in Chapter I

Acrocyanosis —Like Raynaud's phenomenon, this is rather a physical sign or symptom complex than a disease The name merely means blueness of an acral part, almost necessarily an extremity The arteriospasm which causes the blueness is permanent The hands and feet alike—in this case the feet rather more than the hands—are continuously blue, cold, and sweaty Probably in different parts of the world the condition varies in this or that detail Crocq, who named it (1896) described it as occurring in young hysterical women It is certainly rather common in girls, especially Jewish girls, appearing at about the time of puberty Some of these individuals suffer from pes cavus as if there were some slight congenital background such as a spina bifida occulta But the same state or one indistinguishable from it may crop out in persons of any age, men as well as women There is one particularly troublesome form which has been called *Erythrocyanosis Frigida* Telford and Simmons, in their excellent account, say that this is known on the continent as erythrocyanosis crurum puellarum and in France as erythrocyanosis sus malleolaire (above the ankle) Its remarkable feature, as will presently be told, is ulceration of the lower leg which takes a most disabling and intractable course

Acrocyanosis in its mildest form is very familiar, in its serious form, rare Once established, it seldom altogether disappears, though the adolescent sort tends to be outgrown with maturity and in many individuals is only really troublesome in cold weather The degree of redness or blueness varies with the vascularity of the particular skin The change is by no means confined to the digits, for the whole hand is discolored, the whole foot and even the leg for some inches above the



TRAUMATIC EDEMA (REFLEX DYSTROPHY). Following an injury to the left foot without fracture. The atrophy of the bones of the foot (roentgenogram) was extraordinary. The skin of the foot and leg displayed hypesthesia and advanced hypersensitiveness to scratch or pinch. Six hours after a diagnostic spinal anesthesia, the edema had nearly disappeared (Courtesy of Dr. John B Cross, Atlanta)

PLATE IV



ERYTHROCYANOSIS FRIGIDA, A SPECIAL FORM OF ACROCYANOSIS. On the left the preoperative state, the ulcers are open, the skin of the feet and lower leg dark (reddish blue). On the right, the postoperative state, the ulcers healed, the skin of natural color.

PLATE V

ankle. As one passes a hand down the leg from the knee, a coolness is usually encountered perhaps half way to the ankle, a coolness which, in many cases, becomes actual coldness as the ankle is reached. There is very little discomfort, but the cold feeling may be very disagreeable. In damp cold climates, chilblains occur.

Erythrocyanosis Frigida.—The acrocyanosis of young girls may take this serious form. The trouble shows itself chiefly in the feet and legs, overshadowing the moderate blue dampness of the hands. It is always bilateral. Not only the feet but the lower half of the legs are purplish, especially on the posterior surface. In a fair proportion of cases, ulcerations occur. Anywhere between the malleoli and the mid-calf, indurated nodules appear and slowly break down into ulcers. There may be two to a dozen such sores which are of a moderate depth and hardly exceed one cm. in diameter. The lesions are distinctly reminiscent of erythema induratum, or Bazin's disease, but while the latter occurs upon the front of the thin, cold legs of ill-nourished girls and is believed by many to be a form of tuberculosis, erythrocyanosis frigida attacks well-nourished, even fat girls. The ulcers of both, however, are equally chronic and difficult to cure. The diagnosis is not at all difficult and the cyanosis and coldness are readily driven away for the moment by inducing a reactive hyperemia. The pulses are quite normal, the vascular spasm taking place presumably in the arterioles. This is in contrast with the recurring spasms of Raynaud's phenomenon in which the digital arteries are the scene of the vasoconstriction. Plate V, opposite, pictures the case of M. MacL., described on page 151.

Treatment of acrocyanosis need seldom be radical unless the sense of coldness is very troublesome or ulceration occurs. If any deformity of the foot exists, a spina bifida occulta should be looked for but can rarely be treated directly with success. Apparently, like Raynaud's phenomenon, acrocyanosis is aggravated by psychic upsets as well as cold, so that protection in these directions is required. Though the milder cases, if they occur in young girls, will usually become less trouble-

some with advancing years, the serious and especially the ulcerated states should be subjected to lumbar sympathectomy The results of this operation are excellent

The following are instances, respectively, of a mild and of a serious form of the disease

B Y, a Jewish girl, thirteen years of age, complained of cold, blue fingers and toes The catamenia had been established *a year earlier The child* was of unusual intelligence, played the violin, ate her meals in a hurry, and got insufficient sleep Her nutrition was good

For three years, the coldness, dampness, and cyanosis had been increasing in both feet and hands The feet had given the most trouble, apparently because the little toes had progressively overlapped the fourth toes, finally projecting so far dorsally that another surgeon, two months earlier, had amputated both The balls of both feet had become prominent, the arches high, the proximal phalanges extended and the distal joints flexed The child was aware that any emotional strain—her violin lessons, seeing a doctor—made the coldness and sweating worse

The hands were reddish blue in color, cold and damp The change toward normal came rather gradually at the wrist The toes and feet were more blue than red The legs were dusky Coldness was marked up to a rather definite level just above the ankle No changes in the subcutaneous tissues were present and no ulcerations Studies of cutaneous temperature were not made

The patient was advised to eat slowly, rest after her midday meal, secure at least nine hours of sleep and protect her hands and feet from cold When seen two years later, the feet were less troublesome, the hands were about the same Sweating was less noticeable

Seven years later, at the age of twenty, the patient made no more complaint of her feet Both fingers and hands were reddish blue Their palmar surface was glistening wet (when she came in for examination) yet the rest of the body sweat no more than seemed natural The patient, without feeling that

her life was miserable, would have welcomed any operative relief.

M.MacL., a girl, seventeen years of age, complained of recurrent ulcers of both legs. For the last few years, these had broken out with the first snow-storm and had disappeared in the spring. Her feet perspired more readily, she thought, than those of others and frequently felt subjectively cold. She appeared for treatment in March.

She was a good-sized, somewhat obese girl. Examination of all systems and organs was negative save for the legs. The hands were perhaps a little red, but not beyond familiar limits. The blood pressure was 95/60. Both legs, for their lower two-thirds, were mottled and reddish in color. Just above the ankles were half a dozen ulcers on each leg. These were much alike, shallow, round, only a little moist and surrounded by a red tender areola about one to two cm. in width. The feet and toes resembled the legs but were not ulcerated. Both dorsalis pedis arteries pulsated normally.

At a room temperature of 27° C. (80° F.) the temperature of the great toes was exactly that of the air and somewhat lower than that of the skin at the upper margin of the ulcerated area. Here the temperature was about 29° C. (85° F.). Spinal anesthesia brought about a maximal rise upon the great toes, to 34° C. (95° F.) and upon the mid-leg to 32° C. (90° F.). This reaction was considered favorable for relief by bilateral lumbar sympathectomy and the operation was performed transperitoneally. The immediate result was entirely satisfactory. The ulcers promptly healed and the temperature of the skin *remained high*, being little affected by local or general exposure to various environmental conditions. The late result will not be known until several winters have passed.

Hyperidrosis

In describing the various states of vasomotor spasm, damp or even wet hands and feet have usually been described as associated disorders. Sudomotor activity is in fact almost in-

variably associated with sympathetic vasospasm, especially the persistent sort aggravated by cold and emotional stimuli In some cases, the vasospastic blueness and coldness is more and the sweating less, but with the sort of hyperidrosis which is really disabling, the cyanosis is less and the sweating more When the hands and feet sweat excessively they very rarely flush red at the same time, there is almost inevitably some vasoconstriction

Clammy, or wet, hands and feet are more than a nuisance They are actually disabling The slightest nervousness makes the skin glisten and some water actually flows White, in an excellent account, tells of some special cases, of his own and others, of a man who felt he could not practice law because he must so often shake hands, of a medical student whose rubber gloves filled with sweat at the operating table The change is limited rather sharply at the wrist and ankle, and the individual perspires no more over the rest of the body than do others Both sexes are equally affected

Treatment by any other means than sympathectomy is very unsatisfactory Formalin (five per cent) soaks macerate and irritate the skin Radiation, if just the right exposure is used, causes atrophy of the sweat glands at the risk of a chronic dermatitis Sympathectomy is especially suitable because after the sympathetic pathways are blocked, the glands can not be excited to secrete by adrenal influences (apparently they respond only to the chemical action of acetylcholine) Thus, even if the postganglionic neurones should be destroyed, the sweat glands will not be exposed, as are the arterioles, to the action of sympathomimetic hormones

The appropriate operations to denervate the sweat glands of the hands and feet are exactly those advised for vasospastic states and described earlier in this chapter In the thoracic sympathectomy, the stellate ganglion should not be disturbed lest Horner's syndrome set in, and in the lumbar operation it is sufficient to remove the second and third lumbar ganglia and the connecting chain Unless the operator is very expert, the lumbar operations had better be carried out at separate

sittings. The same is even more true for the thoracic sympathectomy. The results should be permanent.

Arterial Embolism

When an embolus plugs an artery it is almost invariably true that it has come from a diseased heart. Thus the background of the most sudden closure to which a great artery can be subjected is unfavorable to life. As a rule, the heart is fibrillating, the left auricle dilated as in mitral *stenosis*, but *sometimes*, because of a coronary infarct, part of a thrombus is detached from the left ventricle. Actually, any dilated heart, in the presence of congestive heart failure, may be a source of arterial embolism. Very rarely indeed, the cardiac disease is so little marked that the source of an embolus can not be surmised.

Emboli tend to lodge where a great artery divides. The bifurcation of the common femoral at the groin is the most common site, nearly forty per cent of all lodgments occurring here. The bifurcation of the common iliac and the aorta between them add about twenty-five per cent more. Thus sixty to seventy per cent of all emboli are more or less accessible from the region of the groin. The popliteal division adds another ten per cent. The rest occur in the arm, principally in the axillary or the bifurcation of the brachial.

If an embolus merely lodged at one of these likely spots, obstructing the vessels here and nowhere else, the result would be bad enough (it has been estimated that perhaps one in five limbs would be lost from a pure occlusion in the common femoral and twice as many from one in the popliteal) but closure of the artery by the embolus is not the whole story: the arterial tree beyond the plug is thrown into spasm and thrombosis is often propagated from the point of obstruction far down the vessel.

A classic [illegible]
a sudden ag[illegible]
and not du[illegible] of the *embolus* itself but to the resulting ischemia. Such an event, coming out of a totally clear

sky, is not, however, invariable Often there are prodromal signs, due either to small warning emboli or to the incomplete obstruction of the artery by the final large one as it is molded in the vessel These signs take the form of a feeling of numbness or of tingling and coldness, the sort of thing one might expect with the onset of thrombosis, and the exact moment at which an embolus becomes lodged is not always easy to fix Pain, however, is the rule and is usually severe enough to indicate the hour from which time elapsed after the *complete* obstruction can be counted There is a great difference between embolism in the leg and in the arm In the latter case, the initial coldness, or it may be pain, is severe, and though the forearm and hand, in embolism, become cadaveric, gangrene of any consequence seldom follows In the former case, the situation is different In middle age and beyond, the arteries of the leg are never as elastic, nor is a collateral circulation so easily established, as in the arm Thus the lodgment of the embolus is followed at once by coldness and a cadaveric appearance of the foot and more or less of the lower leg When the stoppage is such as to have allowed some blood to push, for a time, into the periphery, there is apt to be edema and a good deal of blueness A considerable amount of blood may remain in the small veins This will give a false impression of vascularity, for a pressure spot will quickly become colored again and even a small vein will refill (from the peripheral direction) after being emptied by pressure Seldom is the foot of arterial embolism altogether white It is always more or less cyanotic and if the color be compared with that of the opposite leg, it will usually be clear that the change mounts well above the ankle, in rare cases into the thigh Within an hour or two, the upper level of coldness will also be evident If this is at or above the middle point of the lower leg, the prognosis is poor for survival of the foot If it is merely just above the ankle, the level which marks the upper limit of vasoconstriction in the foot, both foot and leg may well be viable

Bad signs in arterial embolism are, then, coldness, cyanosis

and edema, especially if these mount well toward the knee and if all pulses below that in the common femoral are absent. Good signs are some preservation of pinkness (however faint) and warmth, preferably in the foot, but at least as low as the ankle. The level at which pulsations are felt demands special consideration.

Lodgment of an embolus at the femoral bifurcation obliterates all pulses below the groin, that is, the femoral in Scarpa's triangle and below, the popliteal, posterior tibial, and dorsalis pedis. Just below the inguinal ligament, a *vigorous pulse* will be felt in the common femoral. The artery may even be palpably thickened. Both pulse and thickening may be due to the embolus which transmits a strong arterial beat from above. Lodgment of an embolus at the common iliac bifurcation may or may not completely obliterate the femoral pulsation. However, it always weakens it and sooner or later obliterates it altogether. It is not unheard of for an embolus to catch and then slip down, or, after lodging, to set up a thrombosis, finally plugging the artery after only partly closing it at first.

Lodgment at the aortic bifurcation generally blocks one arterial tree more than the other, but there will usually be evidence of *some* diminution of pulse in both femorals.

In determining the extent to which an arterial pulse extends down the leg, an oscillometer is valuable, but even without it a blood pressure cuff is helpful. If any oscillation can be detected at a particular level, it may of course be due purely to a collateral circulation. However, it is not particularly important, so far as the life of the leg and foot is concerned, to distinguish between a pulse transmitted through the main artery and one derived from collateral vessels. The main thing is to know whether or not a good pulsation is present in the mid-calf.

To bring out some of these points, the condensed histories of several cases of embolism to the lower and upper extremities are quoted from the records of the Peter Bent Brigham Hospital:

M.E.F., Surgical 27614, a woman, thirty-four years of age,

suffering from rheumatic heart disease, in the form of mitral stenosis and aortic insufficiency, had been reasonably well until the onset of the illness which brought her under observation Three days before this moment, she had suddenly been struck down by a violent, almost unbearable pain in the pelvis which radiated down the inside of both legs, especially the left Her vision was blurred, she vomited, and became semi conscious Both feet turned bluish white Her left leg and foot were cold, numb and pulseless Three days later there was still only a feeble pulse in the common femoral Yet nothing worse than a purplish area outside the left heel had appeared. *In the right leg, all pulses had returned In two more days,* an effective collateral circulation must have become established, because pulsation reappeared in the left foot and the patient recovered A rider thrombus evidently caught on the aortic bifurcation, only to slide down into the left femoral The patient's youth and dilatable vessels saved her leg

F J McK, Surgical 59780, a spare man of twenty seven, suffering from rheumatic valvular disease, had been fibrillating *for several days when he was seized with an agonizing* pain in both legs Within fifteen minutes he was seen by the surgeon Pain had then settled in the right leg, which was white, cold and pulseless up to the level of the common femoral at the groin Here a distinct pulsating thickening could be felt Feeble pulses were present in the left foot The patient was groaning in agony Within two hours of the accident, the right femoral bifurcation had been fully exposed and an embolus was found in the common femoral projecting into the superficial branch While all approaches were controlled by soft rubber tubing, the femoral was opened, the embolus milked out from below and extruded with a gush of blood when the current was let in from above Repair was followed by complete restoration of the circulation The moment the current was allowed to flow back into the peripheral vessels—and not before—the pain ceased The operative procedure used is shown in Figure 12, page 160

F V D, Surgical 38637, a man of forty seven, suffering

from a serious mitral lesion, entered the hospital fibrillating. Two days earlier, infarction of the left kidney was thought to have occurred. At three o'clock in the afternoon, he complained of severe pain behind his left knee and of numbness in the leg. That evening, pain and numbness left him but in the early morning, twelve hours after the initial attack, his pain recurred more violently than before and by noon had become excruciating. This was nine hours after the second episode: the left leg was then cold, bluish-white in color, and pulseless up to a point just below the inguinal ligament. In this case again an embolus was removed from the femoral bifurcation, but at this late hour was adherent. A long, soft clot was found attached to its distal end. After removal and repair, a feeble peripheral pulse persisted for only a few hours. Evidently thrombosis occurred and gangrene followed. Embolectomy was performed too late, partly because the early symptoms were misleading.

W.G.B., Surgical 37369, a man, sixty-four years of age and suffering from rheumatic heart disease, entered the hospital fibrillating. Four and a half hours earlier, numbness, which rapidly changed to pain, had attacked his left hand. The arm was held flexed at the elbow, the fingers clawed, cold and white, the arm cyanotic, distal to a point just above the elbow. When the arm was placed at a right angle with the body, a pulse could be felt in the axillary just distal to the edge of the pectoralis major muscle but not beyond.

Exploration, about five hours after lodgment, disclosed a solid whitish embolus, three cm. in length, fixed in the upper brachial artery. To its lower end a filmy red clot, two to three cm. in length, was attached. The artery was washed out with a citrate solution and seemed to be clear. Yet after repair, though a pulse could be felt in the lower brachial [illegible] d

[illegible] d

[illegible] The radial pulse had returned. It must therefore be supposed that some spasm, left after embolectomy, finally disappeared.

The second case is of a less familiar sort but as it demonstrates the good effect of resecting an artery thoroughly obstructed and irritated by an embolus, it is worth quoting

T H W, Surgical 50595, a man, fifty-six years of age, suffered twenty-four hours before presenting himself for treatment a tingling pain and pallor of his left hand There had been no sign of heart disease The hypothenar eminence of the hand was especially painful, the fourth and fifth fingers were "white as a sheet" Below the wrist, the whole left hand was cold, dirty white in color, the finger-nails dusky The patient was so fearful of any contact that he guarded his left hand with his right At first, a radial pulse was palpable but the blood pressure at the elbow was only 110 systolic as compared with a pressure of 170 on the right A peculiar feature was a systolic bruit which could be heard just above the middle of the left clavicle

After nine days of treatment by intermittent use of alternate suction and pressure in the glass chamber, the bruit disappeared and a long tender mass could be felt in the lower course of the radial artery The color of the hand as a whole had improved but the fourth and fifth fingers remained bluish white, clawed, and very sensitive to touch The skin temperature of the left thumb and the tip of the little finger were barely lower than that of the corresponding parts of the right hand but rose only half as much as that of the right hand following a large injection of antityphoid vaccine Some peripheral influence evidently was not only causing vasoconstriction but decidedly checking reactive hyperemia in the left hand The tender, thickened radial artery, giving the impression of an acute arteritis, seemed to be responsible It never occurred to anyone that an embolus had caught in the subclavian and later slipped down into the radial, yet such proved to be the case Resection of the lower radial showed it to contain an embolus and at the moment of resection, under local anesthesia, the patient found himself suddenly relieved of his sense of coldness and sensitiveness in the fourth and fifth fingers The color of the fingers improved and warmth

returned. The vicious circle of arterial irritation and vasoconstriction had been broken up. Subsequently the state of the hand continued to improve.

Treatment.—In spite of failures such as are illustrated by the case of F.V.D., failures which are only too common, embolectomy is still the method of choice in the treatment of embolism in the arteries leading to the legs. The Swedish surgeons, Key and Nystrom, have shown what can be done by concerted effort, but only when patients are promptly brought to the operating table. Unfortunately it is not easy to bring patients suffering from fibrillating hearts and suspected embolism quickly to the surgeon, nor is it always easy to be certain of a diagnosis, even when embolism occurs, as it is very likely to do, in a hospital. One can only hope that by education, better collaboration between physician and surgeon can be secured. It is especially important that hospitals should be equipped with the few simple tools necessary for opening and closing arteries. The fine needles and silk are now furnished prepared and sterilized. Soft rubber tubing is preferred for the purpose of lifting up and checking the flow in a large artery. A long, smooth probe, a fine rubber catheter, through which suction can be made, and one per cent sodium citrate solution or physiologic saline are readily available.

It has been abundantly demonstrated that after more than ten hours an embolus can rarely be successfully removed, because it will have so injured the intima that thrombosis will follow embolectomy, or thrombosis will have spread from the embolus into the peripheral tree. Even a four-hour interval may be too long, yet one hears now and then of success after twenty-four hours.

For the lower limb, attack on most emboli can be made from below the inguinal ligament. The most frequent site is the femoral bifurcation, yet if the embolus is lodged higher, it can still be dislodged by probing upward and by making an extraperitoneal approach through which extraction of the embolus can be aided by massage (Nystrom). By laying bare the common and superficial femoral arteries, through a generous

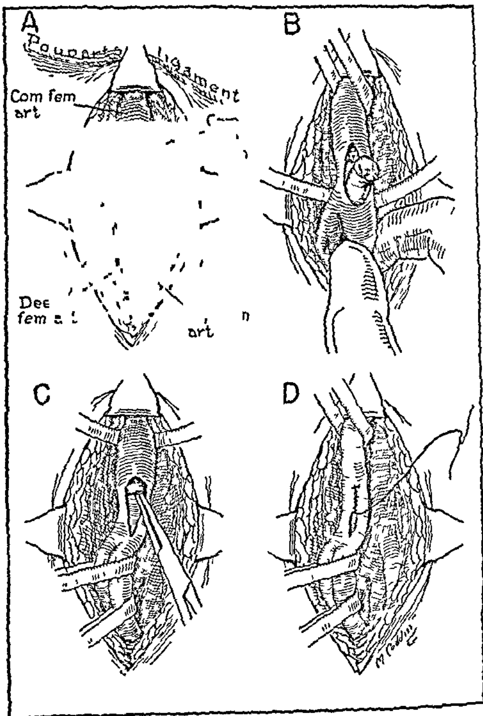


FIGURE 12 ARTERIAL EMBOLECTOMY A diagrammatic sketch after an actual operation *A* The right common and superficial femoral artery are distended by a great embolus *B* The femoral and its branches have fully been cleaned and isolated, and soft tubing has been passed about them The lower part of the thrombus, in the superficial femoral, has been milked out *C* Retrograde bleeding controlled by tubing about the superficial and deep femoral branches While the common femoral is ready to be compressed at any moment by the upper tubing the upper part of the embolus is loosened *D* All bleeding controlled Repair, everting the intima

incision made under novocaine infiltration, the site of the lesion can almost certainly be discovered and further exploration, if necessary, planned. The artery must be handled as little as possible, its intima touched only with the smoothest probe or blunt hook. Suction through a soft catheter and gentle pressure will deliver most emboli once they are looosened. In closing the artery, intima is united to intima with a continuous stitch of fine Chinese silk.

For the arm, embolectomy is seldom required, yet it is, if properly performed, all gain and no loss and may well be attempted in elderly persons in whom a good collateral circulation is unlikely to develop.

For long-standing cases, in which there still seems some hope of saving a limb, resection of the plugged artery offers a decided advantage. For it often relieves vasospasm in the peripheral part of the arterial tree, and may just turn the scale in favor of a nearly gangrenous foot or hand.

Conservative Treatment should invariably be used (1) at an early stage until operative treatment can be secured and (2) when embolectomy is for any reason impossible of accomplishment. It consists in the application of warmth to the body and to the affected limb, slight lowering of the limb, between ten and fifteen degrees below the horizontal, and the injection of such a vasodilator as papaverine hydrochloride. Such measures are fully discussed by Allen and by De Takats.

Heat is applied by placing a *large* cradle over the lower half of the patient's body and legs. The temperature should not exceed 100° F. (38° C.). Such heat, by releasing vasoconstriction, secures the maximum dilatation possible. The leg had better, to conserve its heat and because warming the *body* under the heated cradle is preferable to heating the limb itself, be wrapped in woolen coverings.

Lowering the limb makes entry of the arterial blood easy, but the color of the skin must be studied. The toes (or fingers) should not be made too pale or too cyanotic but, if possible, pink.

Papaverine hydrochloride is given intravenously in a dose

of one-fourth grain (0 015 gm.) dissolved in salt solution. Twice this amount can probably be used with safety and the dose can be repeated

Whether suction and pressure or intermittent venous compression shall be used depends upon whether such apparatus is easily available There is a general feeling that this treatment is unlikely to save a limb which is not benefited by the measures already described It is more likely to help develop a collateral circulation once the immediate crisis is successfully passed

Juvenile Gangrene

This rare and little understood form of gangrene occurs in children Almost any pointed or terminal parts, usually many simultaneously, become necrotic—the nose, ears, toes, fingers, the tips of the elbows, the knee caps But gangrene may also involve a whole limb, or several limbs As a rule, the gangrenous part dries, shrinks and turns black The background is usually an acute febrile infection, occasionally a debilitating illness

From Martin's excellent account of four cases and his study of the literature, one gets the impression that bacteria carried into the blood stream may occasionally attack the wall of one or more large or many small arteries The vessels are sometimes plugged by emboli, sometimes thrombosed, sometimes the seat of arteritis without thrombosis, and on occasion are undoubtedly thrown into a state of violent spasm by thrombosis of their companion veins, a matter more fully dealt with under diseases of the veins It is hardly credible that any peripheral arterial thrombosis or gross embolism in a child can of itself make an obstruction sufficient to cause gangrene, as of a whole limb There must be, in all cases, an element of secondary vasospasm But this vasospasm, if present, is a single episode, and not in any sense recurring, as in the Raynaud's disease or the reactions to cold It must then be supposed that juvenile gangrene can arise under a variety of circumstances utterly unpredictable and generally in the pres

nce of infection; that the arterial or venous occlusion which ccasions it is a source of such local and prolonged vasospasm hat considerable masses of tissue may undergo rapid necrosis, and that repetition of the episode need not occur, provided he basic disease is relieved.

Treatment, being powerless to prevent gangrene, is devoted to curing the infection back of it if such is discovered. That, however, is routine, as in the case of diphtheria, typhoid, or pneumonia. If the child is debilitated and anemic, transfusions are apt to be useful. The gangrenous part must be allowed to separate, the adjacent tissues being encouraged to heal, so that every living bit can later be used to secure, by a plastic operation, the most useful amputation stump, or the least disfiguring remains of an ear or nose.

Erythromelalgia: Erythermalgia

This peculiar and rare symptom-complex is the very opposite of Raynaud's vasoconstriction in response to cold. It is, in fact, a vasodilatation in response to heat. As Weir Mitchell first described it, the symptoms appear in middle life. The *individual first notices* pain in the ball of the foot or toes upon standing, walking, or even letting the legs hang. Soon the dependent part becomes deep red and sensitive to pressure whenever the pain appears. With the deep redness go engorged veins, pulsating arteries and a hot skin. Since that early description, the disease has seemed rather vague and difficult to identify. Most physicians go through life without ever seeing a case, and Smith and Allen, in a recent paper, present only five cases from the Mayo Clinic.

The original name, of Greek derivation, meant red-extremity-pain. Smith and Allen suggest the name "erythermalgia", meaning *red-heat-pain. They find* that the individuals who present the very peculiar symptom-complex are sensitive to heat, that is, in one or more limbs—the feet, hands, or both parts. Once the hand or foot has undergone the violent vasodilatation, the temperature of its skin is found to be elevated to the limit. But the bouts of vasodilatation continue

unchanged for so long as the warmth is maintained Heat the body enough to cause even a slight reflex hyperemia, and as the skin temperature of the part rises to 32° C (90° F), vasodilatation is so accelerated that a temperature of nearly 36° C (97° F) is soon reached Thus 32° C—the point varies slightly from person to person—is a critical level, a sort of trigger point As long as the skin temperature remains in the sensitive range, between 32° to 36° C, the flushing, heat and pain continue The discomfort can even be brought on by artificially inducing venous congestion when the critical level has not quite been reached

The pain comes on as a tingling or pricking, not well localized, but once the hyperemia is full blown, the ball of the foot and tips of the toes, or the corresponding parts of the hand, suffer the unpleasant burning sensation, like a severe sunburn, which Mitchell described To ward off the painful flush, the individual does not hesitate to expose the part to cold, sometimes sleeping with the feet out of bed, going about without shoes on cold floors, and, of course, elevating the feet When the hands or feet are not flushed, there is nothing to observe, though vasoconstriction is sometimes present Polycythemia has been noted in one or two cases

Treatment—Aside from the measures instinctively taken by the resourceful individual to keep the extremities cool, there is little to be done One of Smith and Allen's patients obtained considerable relief from 0 6 grain of acetylsalicylic acid but why the salicylates should be useful is unknown Local nerve divisions have been tried without much success

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CHAPTER V

VARICOSE VEINS

VARICOSE veins are those which have lost the power to transmit blood toward the heart against gravity They are usually dilated, tortuous, and fibrosed Above all, the cup like valves, indispensable to their normal function, are disabled Varicosity is practically confined to the lower extremity and, for reasons which will presently appear, to the parts superficial to the muscular aponeurosis

Varicose veins are totally useless When the body is erect, blood actually flows down them and must be carried off by alternate routes The very fact that in their presence venous blood is still able to return from foot to heart against gravity guarantees the efficiency of those alternate routes Thus, their removal or destruction must always benefit and can never harm the venous circulation For if the efficient veins are able to carry, in addition to their normal load, the down flowing blood of the varicose vein, all the more easily will they function once this unnatural load is removed

Varicose veins, in the dependent position rid themselves of their contents by the aid of two sorts of vessels, namely, the communicating or perforating veins, which pass the stagnant or downward flowing blood through the muscular aponeurosis into the deep system, and the deep veins, which then carry the blood up the leg to the body Thus the communicating veins are a safety vent for the superficial vessels, but the deep veins actually do the work Should the capacious, deep vessels find this something of a burden, the legs feel heavy under exertion and tire easily But should they actually break down, the feet must turn purple and swell Such an event is extraordinarily rare Absence of blueness in the dependent foot of a person suffering from varix is sufficient proof that

the deep veins are functioning normally. If the above is true, some of the modern ritual of tests for varicosity is needless. These categorical statements have an anatomical, physiological, and pathological background which is offered in the following paragraphs.

The Muscular and Valvular Mechanism Governing the Forwarding of Venous Blood from the Legs.—Blood is pushed into the veins of the legs under very low pressure from the capillary bed, the strength of the arterial stream being nearly lost and, when the individual stands, must force its way against gravity to reach the heart. Suction from the thorax can hardly be expected to do more than draw blood toward it from the great abdominal veins. The ability of the legs to empty themselves upwards, under these conditions, is due to their muscular quality and the presence of valves in the veins. The veins are enclosed in what amounts to a muscular envelope, and the valves, bicuspid as a rule, are so placed that blood can flow past them toward the heart but *never* back. In effect, they divide each vein into a long series of segments in such a way that muscular pressure, intermittently applied, permits them to be filled from below and emptied upward.

Not all the veins of the legs are equally exposed to this favorable action. The deep ones, enormously greater in number and capacity, being enclosed within the muscular aponeurosis, are always [illegible] mus- [illegible] walk- [illegible] rficial [illegible] upon them is therefore indirect and is greatly dependent upon the resiliency of the superficial tissues. As years go by and the skin loses its elasticity, the subcutaneous tissue its firmness, muscular pressure becomes less and less effective. Thus the deep veins, well protected at all times, continue to function normally but the superficial veins tend to become distended, to lose a set of valves here and there, and if additional strains are put upon them, fail at last

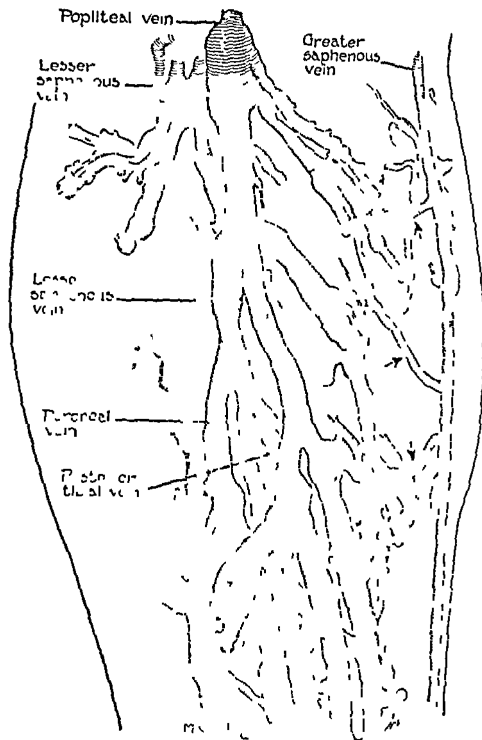


FIGURE 13 THE VEINS OF THE LOWER LEG Sketch after a roentgenogram of an injected leg (presumably at autopsy) Notice the enormously greater capacity of the deep as compared with the superficial veins Arrows point to some of the communicating veins The bones have been removed (Sketch made after Charles Remy's "Traité des Varices", Figure 8 Courtesy of Vigot Freres, Paris)

to transmit blood against gravity. These strains are various and will presently be discussed. The valves require immediate consideration.

The valves are distributed at such intervals that in the great saphenous vein, which drains the median face of the calf and thigh, there are more than a dozen between the foot and groin. There is also one in every entering branch, close to the parent stem. In the perforating veins, which communicate between the superficial veins and the deep system, the valves are usually so set as to allow blood to flow inward but not outward. However, in all communications of this sort the valves permit the most ingenious alternate routes. In time of need, when one part or another of the venous system is obstructed, venous blood is able to pass in a direction never normally taken. The uppermost valve in the veins of the legs is found, rather inconstantly, in the external iliac.

The local arrangement of the valves, as Edwards has shown, is such as to insure their perfect action when muscular pressure is applied to them. Where each pair is attached, the vein is slightly elliptical in cross section, the major axis of this ellipse being parallel to the overlying skin, and each cusp arising from a long side of the ellipse. Thus the crack between the opposing edges of the cusps is likewise parallel to the surface of the limb, and as the skin, or the underlying aponeurosis, presses upon the vein, the latter is flattened and the edges of the cusps are brought together. Blood can of course flow upward past them but is less than ever able to flow back. When only a large single cusp is present, its situation is similarly favorable. Occasionally three cusps are found. Naturally, the valves are dependent upon a healthy state of the vein's wall. They may lose and again regain their competence as the vein becomes overdistended and returns to its natural size. But permanent distention and fibrosis leave them functionless and they are unable to resume their effective shape after thrombophlebitis.

Etiologic Factors in Varix.—Varicosity of veins is perhaps most often due to increased intra-abdominal tension such as

is caused by heavy labor Long hours of standing predispose to it So does pregnancy, but just in what way is not clear, for the superficial veins of the legs may be uncomfortably dilated as early as the second month of pregnancy when increased abdominal tension would seem not to be a factor In other cases, the veins only become distended late in pregnancy, and not until several children have been born is varicosity established Young persons of either sex, girls as a rule, *occasionally begin to notice varicosity at about the time of* puberty, after which the condition becomes progressive Here there appears to be an inborn defect, of valves perhaps, which leads to dilatation of the superficial veins when the legs undergo rapid elongation To some, this type suggests an endocrine influence

Thrombophlebitis *occasionally leads to varicosity* When, as *occasionally happens, the superficial veins, especially those of* the groin and thigh, become engorged as collaterals during obstruction of the external iliac and upper femoral, they are apt to remain dilated and so a completely varicose superficial system results In other cases, the previously normal great saphenous itself becomes thrombosed either independently or in association with a femoral thrombophlebitis Then, its valves being destroyed, it *loses its power to forward blood* against gravity, though it may remain a small, firm, straight cord

Anatomical and Pathological Features of Varix—It is the great saphenous vein and its branches, the principal drainage system of the front and median face of the thigh and leg, which habitually become varicose The lesser saphenous is sometimes *involved through its connections with the greater* but may become varicose when the saphena magna is altogether normal In the latter case, dilated veins are evident upon the back of the calf and external surface of the ankle, close to the heel When the lesser saphenous shares varicosity with the greater, it serves as a *leaking communicating vein, as will* appear below in the description of the tests for varicosity of the superficial and communicating systems

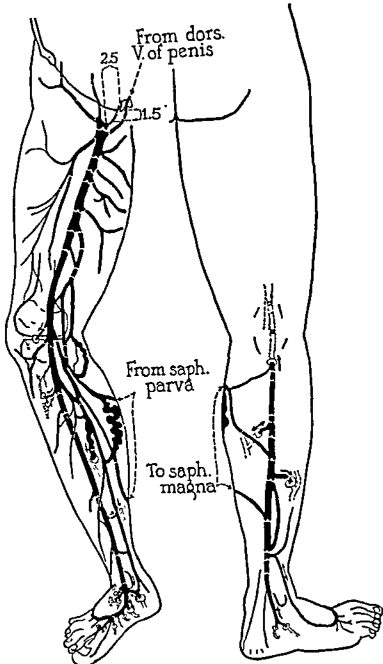


Figure 14. The Great and Lesser Saphenous Systems of Veins, their connections and a few of the communicating veins Note the many branches of the great saphenous close to its entry into the femoral. In the female, some of these would run to the vulva. The valves are accurately shown. From a dissection. Reproduced from Edwards, E. A, "The Treatment of Varicose Veins". Figure 8. *Surg., Gyn and Obst.,* 59.916:928 (Dec.) 1934. Courtesy of *Surgery, Gynecology and Obstetrics.*

The great saphenous vein, as the sketch shows, enters the femoral at the saphenous opening Here it is joined by various superficial branches, some of which come from the lower abdominal wall, some from the pubic region, and some from the thigh itself These vessels must carefully be divided when the saphenous is resected at the saphenous opening For if a varicose stump is left, and these little veins with it, a new varicose system, surprisingly like the old one, is soon organized The great saphenous occasionally splits, in the upper thigh, into two vessels of nearly equal size, but the main stem will always be found to pass toward the median posterior face of the knee Just below this point a rather constant diagonal branch comes off and slants across the shin to the outer face of the calf The principal vein heads straight for the internal malleolus It is in connection with these vessels of the calf that the principal lines of perforating or communicating veins, of which Linton has recently given so full and accurate a description, are found The communicating veins of the thigh are few in number, inconstant, and of little clinical importance

The onset of varicosity is usually consistent with the notion that back pressure causes the saphenous vein to dilate and its valvular mechanism to fail, that is, that varicosity travels from above downward For, though dilated, tortuous veins are first noticeable in the calf, examination of the groin will often show, at the same time, a full, tense, dilated area over the saphenous opening In thin men, such an area forms a visible, rounded lump which transmits an impulse on coughing or straining and is easily mistaken for a femoral hernia In women, the subcutaneous fat is so thick that the dilated saphenous vein of the thigh is seldom visible, though it can usually be traced upward from below by palpation The fact is that in almost every case where permanently dilated veins are visible below the knee, a flow of blood down the great saphenous is obvious, as evidence that the valves in the thigh are gone Sometimes a down flow can only be demonstrated when the individual has long been on his feet, suggesting that *some*

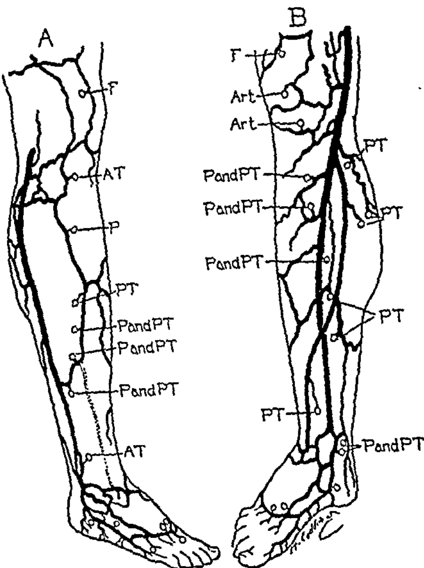


FIGURE 15. THE GREATER, *A*, AND LESSER, *B*, SAPHENOUS SYSTEMS OF THE LEG, showing their communicating branches; after Charles Remy. (Courtesy of Vigot Frères Paris)

The communicating veins are shown by circles and their connections with the deeper vessels (according to Remy) are indicated by letters as follows: *F*, femoral, *Art*, articular; *A T*, anterior tibial; *P*, perineal; *P T*, posterior tibial. It would seem that the communicating veins in *A*, marked *P* and *P T*, should naturally join the anterior tibial or perineal veins, and that those in *B*, marked P and P *T*, should join the posterior tibial exclusively. In any case, this sketch gives a good idea of the number and distribution of these vessels.

valves still function for a time after the vein has, by elevation, recovered its lost tone There is, however, such a thing as local varicosity, if one so chooses to name it That is, a stretch of some visible superficial vein is thin walled and snake-like though never greatly dilated, yet no general varicosity of the great saphenous system is evident Such a state is occasionally seen in multiparous, adipose women and were *it not that a thrombophlebitis sometimes starts in the locally* abnormal vein, would be of little consequence

The pathological change in the wall of the varicose vein is one of fibrosis At first, the vein, though abnormally distensible, is still elastic and capable of contracting Later it becomes permanently dilated, tortuous, and finally hard, even calcified It often gives way here and there, making pockets, some of which reach considerable size but chiefly it takes on a writhing snake like appearance, projecting above the skin in a very obvious manner In thin persons, such a state is almost unmistakable but in the adipose, the skin may be smooth and the dilated state of the veins hardly visible In some cases, the main channel of the great saphenous vein is a single, grossly distended, tortuous cord The branches of such a vein are still quite normal, and the blood it receives is carried off by the communicating vessels In other cases, there are several large varicose branches both in the thigh and calf, but the continuity of the whole venous tree is evident The capacity of such a system is considerable, and one would suppose that the patient, on getting up in the morning, would lose so much blood into it as to become faint, yet one never hears this story Rarely, varicose veins are extraordinarily diffuse, coming to the surface here and there rather than continuously, all over the leg

The state of the skin in most cases of varix is not remarkable Occasionally, pigmented areas appear upon the lower third of the leg, but no sign of malnutrition need be present Certainly, varicose veins lead to no serious degree of edema or cyanosis In other words, the collateral circulation which takes the place of the great saphenous system is usually effi

ient. Ulcers appear to be due to injury, local malnutrition, and infection. They will be considered in a later section. It is astonishing how nearly their location corresponds to that of

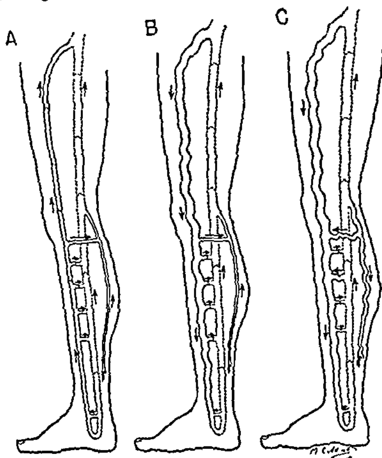


FIGURE 16. *A*. The direction of the current in the normal superficial (solid lines) and deep (dotted lines) veins. *B* Superficial Varicosity. The communicating veins, including the lesser saphenous, are competent and act as safety vents for the varicose veins. *C*. Superficial Varicosity with incompetence of some of the communicating veins, among them, the lesser saphenous.

other ulcers of the leg; that is, in the lower third, more often on the median side than elsewhere, seldom below the ankle, but if so in the region of either malleolus and never on the foot proper. They tend to ride upon a varicose vein. Even when

the area of the ulcer is greatly indurated, a varicose vein can usually be traced down to the site of the sore

The Subjective Symptoms of varix are often remarkably slight It seems to cause more discomfort in the way of tingling or aching when partly developed than when full blown Indeed, well compensated varicosity would be ignored more often than it is if it were not so unsightly, but when varicose veins do cause symptoms the leg is apt to feel heavy on long standing The skin may tingle and often itch quite severely All such troubles are confined to the lower leg as if the degree of back pressure was responsible for them The large veins of the thigh are seldom a source of discomfort, though occasionally, in adipose women, a very tortuous, distensible, superficial vein in this region causes pain Ulceration and thrombosis, after all, chiefly cause the varicose individual to complain

The Diagnosis of Varix Trendelenburg's Tests—Trendelenburg had a very profound understanding of varicose veins, and the simple examinations he devised still afford all the information which anyone requires for diagnosis He showed that varicose veins, once emptied of blood by elevation, fill by a downward rush of blood on depression He understood how the communicating veins are able to carry into the deep system the blood accumulated in the functionless superficial veins The valves of the perforators, he realized, are so set as to favor this flow, but he pointed out that, even when no valves are present, once pressure rises higher in the long varicose column than in the well valved deep vessels, blood must pass from the former into the latter Finally, he devised a test for the efficiency of the communicating veins

Trendelenburg's first test consisted in emptying the varicose veins by elevation, after which the leg was depressed and the patient stood up The blood can usually be seen to flow into the large veins, distending them rapidly from above downward Let this be called a positive Trendelenburg test However, the test is not always easy to carry out, nor is a complete absence of valves invariably evident Varicose veins are often so little visible, or it may be so hard, that the down flow

can not be seen. It can then be detected only by placing the finger-tips upon the empty veins, at a point just below the knee, and noticing their tension as they fill from above. To carry out the test, then, begin by having the patient stand, and view the whole limb, noticing the course and prominence of the veins. Decide whether the down-flow of blood shall be detected by sight or palpation, seat the patient in a very strong chair, and let an assistant tip the chair back. As this is done, elevate the leg to be examined, inspecting, and palpating its empty vessels. Then let the chair be tipped forward and let the patient stand. Should the veins fill with a shock, varicosity is evident enough. If they only fill in five to ten seconds, some valves are still present and are able, until distension of the vessel is complete, to delay the downward flow. Such an event is of no great importance; varicosity may be counted present.

That blood actually flows down a valveless vein when the leg is dependent requires little proof. Without valves, blood could never mount against gravity. But actual proof has been secured by McPheeters and others that blood descends a varicose vein, passes through communicating vessels into the deep system, and there mounts toward the heart. That is, opaque material has been injected into a varicose vein and traced by the aid of the X ray along the course described above.

Next comes the *Trendelenburg Test with Constriction.* Blood is known to flow down the valveless vein. The question then is: can blood still flow into the varicose veins of the lower leg when prevented from flowing down from above? In fact, do the communicating veins leak, allowing blood to escape from the deep into the superficial system? To test this point, the patient is again seated in a chair, and tipped back, the leg raised and emptied of blood. But now carry a piece of bandage about the mid-thigh, passing its ends through the fingers just as reins are, or used to be, held in driving a horse. A twist of the hand tightens the bandage and the patient stands. Again the varicose veins below the knee must be palpated if their state of emptiness or distension is not plainly visible. The constriction indents the thigh sufficiently to prevent a

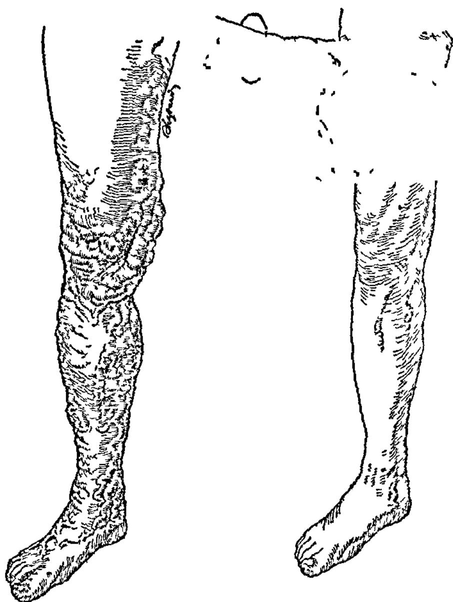


FIGURE 17 THE TRENDELENBURG TEST, WITH CONSTRICTION The leg, having been emptied of blood by elevation, is subjected to constriction in the mid thigh by the tightening of a piece of bandage In purely superficial varicosity, down flow of blood is prevented and the superficial veins, below the constriction, do not fill—for half a minute or more Even when moderately full, they become increasingly tense on release of the constriction In the presence of incompetent communicating veins, filling occurs rapidly below the constriction and there is little or no additional impulse on release of the bandage (From Homans's *Textbook of Surgery* Courtesy of C C Thomas, Springfield, Ill , and Baltimore, Md)

downward flow in the varicose veins but does not interfere* with the circulation beneath the muscular aponeurosis. Should the varicose veins remain empty and relaxed below the constriction for half a minute to a minute (depending upon their capacity; for they must finally be filled from the arterial side) the test may be considered negative; that is, the communicating veins are competent and no blood escapes through them from the deep to the superficial system. This being determined, what the French have called the "contre-épreuve" of the Trendelenburg test may now be completed: the bandage is released and the impact of the down-flowing blood into the varices below is seen or felt.

A positive constriction test leads to further discoveries. Suppose the varicose veins fill below the constriction in five, ten, or fifteen seconds, proving that some communicating veins are incompetent. It may be desirable, especially if a varicose ulcer is present, to discover at about what level the leak or leaks occur. A varicose lesser saphenous vein, which has connections with the greater and empties into the popliteal, should first be suspected. So apply the constriction to the *elevated leg* just below the patella and repeat the test. If the veins now *fail* to fill below the constriction the problem is solved. The lesser saphenous vein is almost certainly varicose and connects with the varicose great saphenous system—blood leaks out through it from the popliteal into the superficial veins. Therefore, to free the superficial parts from venous stasis the lesser as well as the greater saphenous vein must be divided, obliterated, or removed. Beyond this it is not easy to go. If the superficial veins still fill, on depressing the leg, when the constriction is applied at the knee, it is only possible to say that incompetent perforators are present at a still lower level.

* There is a good reason for using a bandage rather than a piece of rubber tubing. The latter is so yielding that it fails, until drawn very tight, to shut off completely the downward flow in the superficial veins; and when drawn tight is very likely to interfere with the venous return beneath the aponeurosis, making the foot cyanotic. Whereas it is very easy to twist the bandage just tightly enough to prevent any flow down the varicose veins without causing any deep venous congestion whatever.

However, one may sometimes learn a little more One may wish to know, for instance, where most of the communicating veins are situated So the leg is depressed until the varicose veins are full and tense Then, with the finger tips, one compresses the great saphenous at the groin and elevates the leg, say to an angle of 20° above the horizontal As a rule, the veins of the thigh remain full, there being no perforators (or insignificant ones only) above the knee Below the knee, the superficial veins will usually empty themselves through perforating veins into the deep system, so that one can detect the level of the highest (cephalad) of the latter One can then, with one's free hand, sweep the blood from the thigh toward the foot and note at what point most of it disappears from the surface vessels In the jargon of varix, a positive constriction test—filling of a varicose vein below the constriction—is called a "Trendelenburg double"

The Schwartz Test—In 1908, Chevrier published a monograph on varix in which he called attention to the "Signe de la Chiquenaude de Schwartz" With the leg horizontal, the full vein was tapped in the thigh and the consequent undulation traced toward the periphery, thus demonstrating the absence of valves The writer (1916–27) having adopted this test to his own purposes, used it in the erect position, making the wave travel from below upward and using it to trace the course of the varicose vein in the adipose thigh To this end, the varicose vein below the knee is snapped with the back of the finger-tip and the shock is felt by the flat of the fingers of the other hand placed upon the inner face of the thigh Only a varicose vein will transmit such an impulse Thus the sign is in a way a test for varicosity

Others have observed the same phenomenon

The Perthes Test—A number of clinicians, whose opinion deserves respect, make use of this test Its object is to prove, or disprove, that in the presence of superficial varicosity the deep system of veins is functioning normally Perthes noticed that after Trendelenburg's division of the varicose saphenous vein in the thigh or when the vein was compressed high with

the fingers, exercise caused the calf to become smaller, by which he judged that the pumping action of the muscles was emptying the leg of blood effectively through the femoral (deep) system. On the other hand, if the blood were free to pour down the varicose vein, the calf failed to shrink and even became enlarged, the task of the communicating and deep veins being one of cleaning, as he said, the Augean stables! His compression of the great saphenous at the groin was actually a test before operation of how much more effectively the deep veins would empty the leg of blood after the down-flow through the varicose saphenous vein was cut off than before. However, he did not propose high compression of the varicose great saphenous vein either as a test of obstruction or efficiency in the deep venous system. It is others, in subsequent years, who have held it to be available for this purpose, and recently Mahorner and Ochsner have used it to bring out some rather fine points in diagnosis. Tying *a piece of rubber tubing about the thigh, they set the patient to walking and notice whether the varicose veins, below the constriction, shrink* or remain full. If the veins shrink, the deep veins are effectively draining the varicose system and are themselves patent and efficient. By applying the elastic constriction at various levels in the thigh, they hold that they can identify a point at which the deep veins are obstructed or at which a leaking perforating vein is present. The test ignores the principle already laid down earlier in this chapter, namely, that merely walking or standing, with no constriction applied, tells the story of the deep veins in the presence of varix. For since the deep vessels must *always* do the work of the varicose veins, beside their own, their efficiency is already tested and proved if the foot of the varicose leg does not become cyanotic on walking and standing. One can compare the varicose leg with the normal one, or two varicose legs with those of an otherwise comparable individual. Finally, it should be pointed out that the right degree of constriction with a piece of rubber tubing—improperly called a tourniquet—is difficult to secure. In fact, the Perthes test, with its variation, is one for

experts Skillfully used, it secures information about the level of incompetent perforating veins The presence or absence of valvelessness or obstruction in the femoral system is evident without its aid

Varicose Ulcer—Ulcer is the most common complication of varix and is chiefly responsible for the disablements due to that disease It has already been explained that the varicose leg often presents, throughout life, a healthy skin The change most frequently seen is a deposit of brown pigment, first as a local speckling, later as a deep brown patch Such may precede ulceration The change seems to indicate malnutrition and to appear most often in the lower part of the leg, in front or upon the inner surface, occasionally over or below the malleoli

Ulcers are apt to appear in pigmented areas and on the course of veins, that is, they tend to "ride" veins A trifling injury usually initiates the process The first sore is trifling, shallow, and as a rule is readily healed by cleanliness and the use of a bandage sufficiently firm to compress the veins to which the ulcer is tributary But a spot once ulcerated, tends sooner or later to break down again In time, the sore enlarges and becomes the center of an area of more or less marked induration and edema Just how varicosity lowers the resistance of the tissues, is immaterial Local malnutrition, followed by injury and infection, is sufficient excuse In some instances, however, the location of a sore is dictated by a great lake of varicose vessels or by the presence of a leaking communicating vein beneath This latter possibility needs keeping in mind, for if a leaking perforator is present, merely doing away with the great saphenous vein proximal to the ulcer will rarely cure it, local venous stasis being maintained by the incompetent communicating vein This is the best reason for using the constriction test to detect the presence of such veins It may call for special operative treatment to rid the leg of them

Ulcers are more or less painful and disabling according as they are or are not inflamed But even a badly infected ulcer,

as indicated by the redness, swelling and tenderness about it, is more painful in certain situations than others. The worst are low down upon the inner face of the leg overlying the internal malleolus. Perhaps they derive their painful quality from the presence, beneath them, of the long internal saphenous nerve, which is closely associated with the great sa-

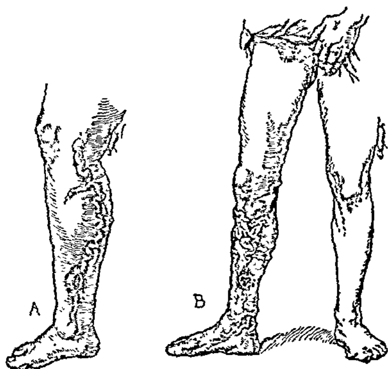


FIGURE 18. VARICOSE VEINS AND ULCERS. *A*. The ulcer "rides" upon a vein *B* The ulcer is in the midst of a considerable induration and is less directly related to any one varicose vein. At the groin, a dilatation over the root of the great saphenous vein is shown. (From Homans's Textbook of *Surgery* Courtesy of C. C. Thomas, Springfield, Ill., and Baltimore, Md.)

phenous vein in the calf and supplies the skin of the median face of the leg and ankle. It will sometimes be found advantageous in treating the ulcer to divide this nerve near the knee.

The favorite situation of varicose ulcer does little to distinguish it from other sorts. The postphlebitic ulcer is most

often found in similar locations, and even the unexplained ulcers of the adipose are usually seen on the median face of the leg in its lower third Nor does the appearance of a varicose ulcer mark it in any way It is usually shallow and not undermining, but so are most other ulcers (except syphilitic ones) It rarely encircles the leg but others rarely do so either In fact, a varicose ulcer is identified principally by a history of, and by the presence of, varicose veins Destroy the varicose veins and some varicose ulcers will persist, either because of great thickening and scarring of the surrounding tissues or because of incompetent communicating veins beneath But just because an ulcer *looks* like a varicose one, it should not so be labeled, and treated, unless varicose veins are plainly responsible In these days, when a multitude of clinicians seem to be in the habit of filling any vein they can find with a sclerosing solution just because an ulcer is present in the lower leg, this prohibition is important Many ulcers so treated are postphlebitic ones and are made worse by such thoughtless acts The postphlebitic ulcer will subsequently be discussed as a separate problem

"Varicose Eczema"—In a small percentage of cases, a patch of dermatitis, usually described as "eczema", is associated with varicose veins This is most often situated on the inner face of the lower half of the leg, just as is the case with ulcer, but may appear upon the outer surface or even occupy the whole lower leg The patch is slightly elevated, reddened, scaly, and constantly weeps, but is not actually ulcerated Plate VI, facing page 202, illustrates an advanced but perfectly typical disease of this sort Apparently varicose veins merely occasion the change which favors the establishment of such lesions, and it is almost equally probable that all these dermatoses, if such they may be called, are not of one sort However, since signs of fungus infection on the feet—scaling skin, moist, itching patches between the toes—almost invariably are present, it may be supposed that most of them are allergic reactions to, if not the immediate seat of, epidermophytosis

Treatment is exceedingly difficult. In the first place, the varicose veins had better be divided at the knee and groin. Even if incompetent perforating veins suggest the need of a dissection of the calf, this will hardly be possible in the presence of an infected skin. Probably life in bed for a week or more, while gentle liquid fungicides, such as potassium permanganate (1 to 2000-3000), thymol or salicylic acid in fifty per cent alcohol (1-2%) or watery aluminum acetate (1 to 12-20) are applied to the leg and foot, should precede the high-low division of the saphenous vein. Any considerable dissection must be postponed till later.

In the subsequent treatment of the "eczema", the sensitiveness of the patient to fungi as well as other possible causes of an allergic reaction should be studied. Any *recognizable epidermophytosis* should be treated by fungicides carried in solution or ointment as trial directs.

Treatment of Varicose Veins and Ulcer

Nonoperative Treatment.—This comprises the application of bandages or stockings and the injection of sclerosing chemicals.

The principle upon which *Bandages and Stockings* are applied is a very simple one, namely, that the blood which otherwise would *fill*, *distend*, and even flow *down varicose* veins is by the pressure of the bandage *prevented* from so doing. The varicose veins are *compressed*, though one can hardly expect them to be kept altogether *empty*, and the venous blood, instead of remaining *stagnant* in the superficial parts, is assisted by the elastic pressure to enter the deep veins which are quite able to care for it. The modern semi-elastic cotton bandage is very effective, as is the *elastic* stocking, but an unyielding bandage, if carefully *fitted*, serves almost equally well and can be left on for many days. A lace-up canvas stocking is also very useful.

In the presence of *ulcer*, such palliative treatment is intended: (1) to keep stagnant blood away from the sore and (2) to aid in applying such local ointments or antiseptic solu-

tions as will diminish infection and favor epithelization When afflicted with large, infected, and deeply indurated ulcers, the patient had better be confined to bed, the leg slightly elevated Hot saline or boracic dressings should then be applied A dehydrating solution such as glycerine and saturated magnesium sulphate, if not too painful, may be of advantage As edema diminishes, the ulcer will take on a bright color and skin will begin to cover it Such treatment must usually precede one of the operative measures later to be described Even if it induces healing of the ulcer it can not be expected to produce a permanent cure

For ambulatory treatment of a difficult ulcer, the use of the rubber sponge, or "venous heart" as described by McPheeters, is very effective "A good grade rubber bath sponge is selected of a size larger than the ulcer Some soothing ointment is applied to the ulcer surface Fluffy gauze dressings are applied and a few layers of sheet wadding Over this the rubber sponge is applied and bound in place with a plain gauze bandage * * * Now apply the four inch ace cotton elastic bandage starting at the knee and going downward with a double figure of eight about the ankle " Walking is then encouraged as it tends to pump fluid out of the tissues

Another excellent ambulatory dressing, seldom used because its application requires time and pains, is the Unna's paste* stocking This is indicated when there is little infection but the ulcer is resistant to epithelization The application of the stocking requires some skill The leg, after being elevated for half an hour, is painted thoroughly with the warm liquid paste At once a single layer of narrow gauze bandage is applied from toes to knee, over skin and ulcer alike Upon this a second painting is made Then another layer of bandage In this way three or four alternating layers of paste and gauze are applied until the bandage is fairly firm though still flexible If the ulcer is relatively clean, such a boot is left on for even a couple of weeks, at the end of which time healing

* To make Unna's paste mix zinc oxide 10 gm. gelatin 40 gm., glycerine 120 ccm. water, 150 ccm heat in a water bath to liquefy before applying

will often be complete. Or a window can be cut for dressings. As a substitute for this boot, a very useful procedure, in the case of a clean, shallow ulcer, is to cover it with strips of zinc oxide plaster, which half encircle the leg and make a little pressure upon the sore. Over this, even without an intervening dressing, a bandage (from toes to knee) is applied.

The nature of the local application seems on the whole the least important aspect of treatment. In other words, vaseline gauze is about as efficient as boracic ointment and boracic ointment almost as good as an ointment * of oxyquinoline and scarlet red. Sometimes a sore can be treated effectively by regarding it as a burn and painting it with a dye such as gentian violet (2 per cent), to form a dry covering, or with a 10 per cent solution of silver nitrate.

The Injection of Sclerosing Chemicals.—This method has now been long enough in general use so that its virtues and failings can be fairly well evaluated. It is not a cure-all for the lazy surgeon to use. Indeed, the successful injection of sclerosing fluids into varicose veins calls for a high *degree of skill* and pains. It is probably true that as the great majority of surgeons practice injection, they fail to obliterate permanently more than a small percentage of the veins they treat, which is not to deny that such injection may here and there close a short length of vein. However, their injections, by at least *temporarily obliterating* one or more veins leading to an ulcer, will cause it to heal while the patient is ambulatory, a most important consideration for those who must work daily. Two or perhaps three serious indictments can be brought against the injection method: (1) In unpredictable instances, a disagreeable, wandering, obstinate sort of thrombophlebitis is set up; (2) recurrences are common and often so diffuse as to be *difficult to treat* by any method, and (3) it is not easy to avoid an occasional slough.

	Gms. or Ccm
* Oxyquinoline sulphate	0.6
Scharlach R ointment 5%	120
Liquid petrolatum	15.

The first two undesirable happenings just described can be avoided, or rather minimized, by combining with the injection a high division of the great saphenous vein, that is, at its junction with the femoral A retrograde injection made during this procedure is seldom followed by any serious degree of soreness of the injected veins and the combination certainly lessens enormously (over pure injection) the number of recurrences However, high division, except in very favorable cases (thin men) requires hospitalization for forty eight hours It is not a routine procedure for the outpatient department or office

To these general criticisms it should be added that very large veins are difficult to close by injection, and that injection is particularly ineffective in the presence of leaking communicating veins (doubly positive Trendelenburg test) In the treatment of postphlebitic ulcer, injections are utterly useless and even dangerous, not because they may not obliterate a dilated vein now and then, but because they aggravate the already inflamed state of the tissues, increasing disability and pain and even causing new ulcers to appear

The indications for injection, pure and simple, that is, without high division, are, first, the treatment of varicose ulcer when the connection between vein and ulcer is clear, especially in working people and the aged (in the former it should, however, be followed up by more radical treatment) and second, the obliteration of unsightly but otherwise symptomless varices of moderate size

High division plus retrograde or secondary injection is indicated in the treatment of any varices at any age, provided that incompetent communicating veins are not present (indication for open resection) or that a deep thrombophlebitis has not left behind postphlebitic indurations

To the above should be added two general rules, which, if religiously followed, greatly favor the success of injection (1) Inject as great a length of vein as possible at one sitting, and (2) inject the vein when empty of blood, keep it empty for a few minutes afterward, and, by compression, prevent it

from being distended with blood during the following twenty-four hours.

Injection without high division.—The length of vein to be treated should carefully be studied. It may require the use of several needles, a matter calling for a certain degree of quickness and skill—one needle perhaps to a stretch of 7.5 cm. If the veins of both thigh and leg are to be treated, McPheeters recommends that the thigh be treated at the first sitting, the leg at the second. An encircling piece of rubber tubing prevents the sclerosing solution from passing down from the vessels of the thigh into those of the calf. The needles used should have a short bevel and should not be too fine lest blood clot in them before the injection can be made. Each is inserted attached to an empty syringe, while the vein is reasonably full of blood, the leg slightly dependent. Blood must then be demonstrated by suction, after which the syringe is detached and the needle, from which a little blood oozes, left in the vein. If only one injection is to be made, the leg is then raised and, to keep the vein empty of blood, a ring of lead wire is pressed down upon it, isolating the stretch of vein to be treated (or pieces of bandage or tubing are tied above and below the area). If several injections are to be made in series, an assistant may hold a piece of gauze about the open dripping base of each needle until the operator is ready to make his injection into them in turn. The injection is of course made when the leg is elevated. A long segment of vein can be isolated and kept empty by encircling rubber tubing.

For large injections, solutions of invert sugar (seventy-five per cent), dextrose (fifty per cent), or sodium chloride (ten to twenty per cent) are suitable. Such are now available in sterile ampoules; also various favored combinations. As much as twenty ccm. of one of these solutions can be injected at one time. For injection into short stretches, a useful solution is quinine hydrochloride (gm. 0.26) and urethane (gm. 0.13) combined in a two ccm. ampoule. No more than this should be used at one sitting (vomiting may occur if the patient's tolerance for quinine is low). Another effective one is sodium

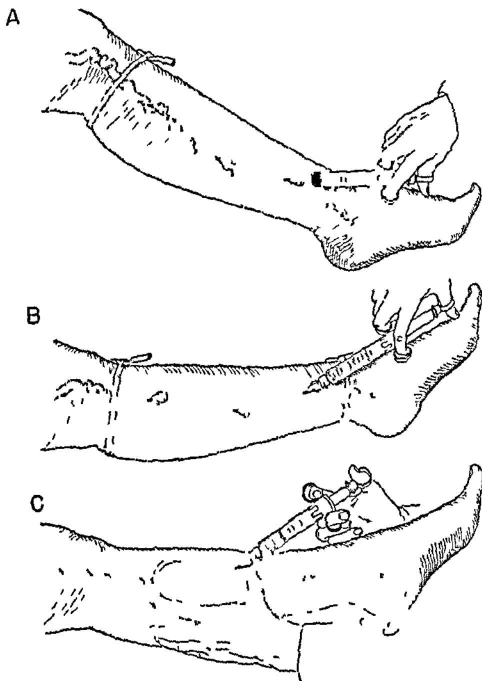


FIGURE 19 THE INJECTION OF VARICOSE VEINS *A* Needles are inserted while the leg is partly dependent and are left detached as successive ones are placed, a rubber band prevents down flow of blood in the vein *B* The leg is quickly raised to the horizontal and the injection made, between bands at knee and ankle, into empty vessels *C* The u e of lead wire to localize an injection, as of quinine and urethane, or sodium morrhuate

morrhuate, in five per cent solution, of which two to five ccm. may be used.

In former times, much sodium salicylate was injected in a thirty per cent solution (or even stronger). This, like strong sodium chloride, causes sufficient pain to call for a general anesthetic and is not now favored.

Following injection, the needle is left in place for a minute while the solution diffuses. As it is withdrawn, pressure is made over the spot with a piece of gauze. A pad is then applied over the injected area and held in place by a bandage. Undoubtedly the efficiency of the injection is increased if the leg is not lowered for twenty-four hours, though active exercise, immediately after the injection, has been advised to carry any excess of solution out of the deep veins. Just why this is proposed when moderate elevation drains the leg still more rapidly, is not clear to the writer.

High Resection of the Great Saphenous Vein.—The idea of combining the injection of a sclerosing solution with ligation of the great saphenous vein in the thigh is an old one, and De Takats seems to have been the first to use a reasonably high division in ambulatory patients for this purpose. The writer prefers to speak of "resection" and that at the highest point possible, the saphenous opening. The word, "resection" is advisedly used in place of "ligation". A vein ligated in continuity re-establishes its channel with astonishing rapidity. Simple division between ligatures is hardly more effective. But division of the varicose great saphenous vein at its junction with the femoral, leaving no varicose stump and removing several centimeters of the vein below, is reasonably effective. Particular attention should be given to the branches entering the vein near its root. As these are divided they should be followed gently into the fat with the point of a fine hemostat and there ligated. It is easy to understand that if a varicose stump is left, with branches emptying into it, connections are soon made with neighboring veins and a new varicose system is soon established. To bear out and expand this statement, the writer has observed that among women

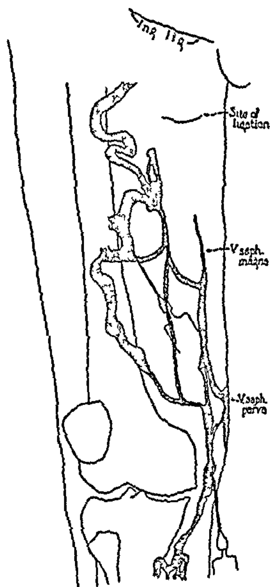


FIGURE 20 RECURRENCE OF VARICOSITY AFTER AN INSUFFICIENTLY HIGH SAPHENOUS DIVISION The new varicose connection might well have been shown passing into the stump of the great saphenous or the vulval veins After an actual dissection of Dr E A Edwards, who has kindly allowed the use of his sketch (Courtesy of *Surgery, Gynecology and Obstetrics*, 59 916 928 (Dec) 1934)

who have borne children and in whom large vulval vessels connecting with veins in the upper thigh are evident, it is a practical impossibility, by any method, to prevent a recurrence of varicose veins. One may resect the upper great saphenous with its entering branches, carry the incision medially close to the vulva, dividing all veins encountered, and make a retrograde injection of the main stem of the great saphenous (or even remove it down to the knee), yet, as a rule, a new set of varicose veins will be established within six months to a year. The explanation seems to be that many small potentially dilatable veins cross the operative field. Mere division of these fails to prevent their making some sort of connection across or around the reunited surfaces. Back pressure from above, there being no valves or only useless valves in the neighboring veins, soon opens up a new varicose pathway.

The technique of resecting the saphena magna at the groin is sufficiently illustrated in the accompanying sketches. The operation is performed under procaine infiltration. The one per cent solution is generously infiltrated into the skin, subcutaneous tissue, and, if the patient is not so fat as to make the whereabouts of the needle point uncertain, into the fascia lata. The operator then waits twenty minutes (by the clock). The vein is necessarily encountered by the oblique cut and once found is then and there isolated and divided between hemostats. It is then dissected upward until the operator can see the bend of the vessel as it enters the femoral, which he must be careful not to draw out. In this part of the procedure he will sometimes be aware of a vague group of lymph nodes, which may take the form of a faintly inflammatory thickening, rather lateral to the varicose stump. He should disturb these as little as possible or an occasional reaction will result, causing some local induration and swelling in the region of the wound. As the stump is lifted up, its entering branches are clamped, followed into the fat and tied with fine silk. Clamps on the stump's side are rarely needed. As the great saphenous itself is ligated (with larger silk), tension on the stump is released

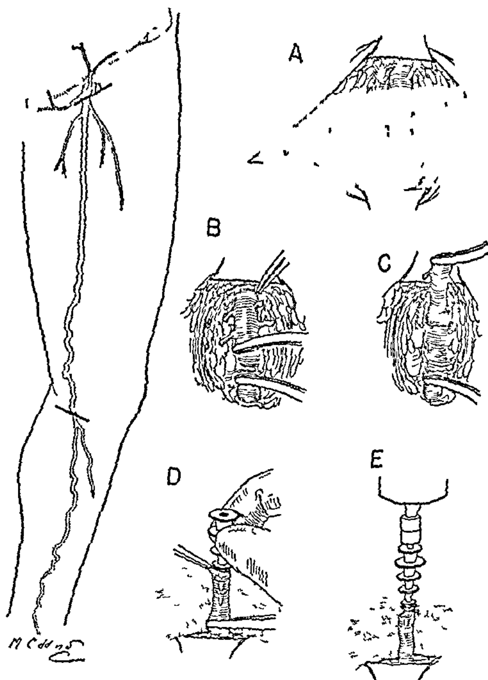


FIGURE 21 HIGH DIVISION (RESECTION) OF THE VARICOSE GREAT SAPHENOUS VEIN The low incision for the 'High Low Division' is also indicated *A* The great saphenous and its highest branches *B* The varicose vein is divided well below the saphenous opening Its branches are followed into the fat, divided and ligated with fine silk. *C* The stump is turned up, ready to tie at its entry into the femoral *D* and *E* The insertion of the Mass Gen. Hospital "pagoda" into the distal stump, ready for injection of the sclerosing solution

to make certain that the femoral is not being drawn out and included in the ligature. A long cuff (1.5 cm. or half an inch) is left distal to the tie. The pessimist will apply two ties.

If a retrograde injection is to be made, it will be of advantage to have operated with the lower end of the table elevated perhaps six inches above the head. The veins will then be empty. By tying a cannula, which fits a twenty ccm. syringe, into the lower end of the divided vein, the fluid is easily injected without leakage. Any of the concentrated sugar solutions, combined with twenty per cent saline, can be used in an amount up to twenty ccm., depending upon the size of the varicose veins. There will often be a momentary cramp-like pain and, after the lower stump has been ligated with silk, the wound is irrigated with warm physiologic saline and closed with fine silk stitches so placed as to obliterate all dead space. A small local flexible dressing is then sufficient for the wound, but a gauze pad may be applied to the inner face of the thigh (to compress the injected veins) and held in place by adhesive strips or a bandage. How long the patient shall remain in bed depends upon the healing of the wound. The leg can be moved freely from the start. If the wound is *reactionless*, one or at the most three days in bed is sufficient, but during this time the patient should remain prone, not sit up or recline. Whether he can then get about without discomfort will depend upon the reaction in the varicose veins. As a rule, a reasonably normal life can at once be resumed. For a reliable account of the ultimate success of this procedure, especially as compared with injection alone, Faxon and Barrow's paper from the circulatory clinic at the Massachusetts General Hospital should be consulted.

Combined High-Low Resection.—An advantageous variation upon high resection with retrograde injection, especially if the varicose veins are large and present one main channel at the knee, is to begin by dividing this vein—the foot of the table being raised six inches above the head—through a transverse incision just below (or above) the knee joint. In that case, any branch connecting with the lesser saphenous vein

can be cut off and the solution, later to be injected from above, will not run down and set up a useless, annoying secondary thrombosis in the calf Needless to say, the division and ligation of all veins encountered must be painstaking and complete, lest a leak of the sclerosing solution into the tissues occur Following this operation, and at the same sitting, the resection at the groin and retrograde injection are carried out The high-low procedure is probably more effective than high resection alone

Operative Resection of Varicose Veins—This operation, nearly discarded at one time in favor of injection, has a place under certain conditions That it was a cause, in persons over fifty-five years of age, of just as much fatal pulmonary embolism as occurs in such individuals following *any* operation upon the abdomen or below is reasonably certain That it was a cause of embolism, in younger persons, unless carelessly performed, is extremely doubtful It may, in fact, be undertaken without hesitation, provided the operator is not planning to hurry over it, in any one, preferably in the twenties or thirties, who desires a radical cure of his varicosity However, it is distinctly indicated (1) when, in a young or middle aged individual, the varicose veins are especially large, (2) when, in such an individual, a varicose ulcer is threatened or present, and (3) when incompetent communicating veins are proved to be a factor, especially in the presence of an ulcer It may, in fact, be elected by patients in either of the first two of these three categories and is more or less mandatory in the last The operation calls for eight to ten days in bed and perhaps two weeks in hospital It requires a perfection of technique and gentle handling of tissues to secure ideal healing of a number of wounds, the least possible immobilization in bed, and unnoticeable scars

The operation is performed under a spinal or gaseous anesthesia, the foot of the operating table elevated perhaps six inches above the head The same technique is employed for dividing the great saphenous vein at its junction with the femoral as is used in a high resection—an oblique incision,

parallel to the inguinal ligament, resection of all branches entering the upper stump, and ligation exactly upon the femoral vein. From its point of division to a point just below the knee, the great saphenous vein may be removed by any convenient method—the Mayo stripper, for instance. The operator may elect to tear the main saphenous stem from its branches and control hemorrhage by pressure, but he should prefer the more surgical method of cutting down upon the vein whenever a group of entering branches is demonstrable. In any case, he should keep in mind that the most important part of the operation is the abolition of back pressure from above by thorough eradication of the great saphenous vein from groin to knee. Here, with fresh instruments,* *the dissection of the lower leg is begun.* As a rule, the course of the principal varicose channels in the calf should be studied and a liberal incision made which will permit their removal without lifting flaps for more than an inch to one and a half inches in a lateral direction. Unless for some special reason, the veins need seldom be removed for more than two-thirds of the way from knee to ankle; if a wide dissection in the lower part of this area seems desirable, a transverse cut, making the whole incision take the form of an inverted T or broad Y, is wise. Since the arterial supply for the superficial tissues of the front and sides of the calf comes from longitudinal rows of small arteries emerging from the deep fascia at quite regular intervals, very wide flaps are likely to slough.

In making an incision for the removal of varicose veins, the dissection should at *once* be carried to the deep fascia (whether or not in so doing the veins are divided in several places). Then flaps of full thickness, including all fat and subcutaneous tissue, are turned up, and from the inner surface of the flaps the veins are removed. This step causes the least possible traumatism to the cutaneous edges and tissues in general, and favors healing without necrosis of the skin.

* There is little more justification for using the same dissecting instruments for a succession of incisions in one patient than for using the same ones for patient after patient.

If a probable connection with the varicose lesser saphenous vein has been demonstrated—that is, a leaking communicating vein in the region of the popliteal space—a *transverse* incision should be made behind the knee The lesser saphenous will usually be found without difficulty just beneath the thin deep fascia, passing upward in the middle of the space No more of it than can easily be reached through the transverse incision need be removed

The Operative Removal of Varicose Veins in the Presence of Ulcer—It is desirable that an ulcer should be clean if not healed before a radical operation is undertaken, since the lymphatics encountered in the dissection may, in the presence of dirty ulcers, become a source of infection

The veins are removed as usual and the dissection is carried as near the ulcer as is considered safe It is convenient to end the linear incision in a very broad inverted Y If the ulcer is of moderate size and little indurated, this will be sufficient But if the ulcer is old and much indurated, it may be excised at the same time with, or some days subsequent to, removal of the veins In the former case, the excision of veins should first be finished and the wounds closed Then the ulcer, with a margin of sound tissue *and the aponeurosis beneath* should be removed in one block, laying bare muscle and, if necessary, tendon sheath, periosteum, or the capsule of the ankle joint The clean surface thus left can at once be covered with an Ollier-Thiersch graft which usually heals without difficulty Such a radical procedure is seldom necessary Most ulcers can be given a trial of excision of the veins alone If that fails, it is time enough to remove them

After-Treatment—When the patient is first allowed out of bed—some ten days after the operation—the leg is bandaged from toes to knee Moderate exercise is encouraged but when the leg is not in use, it is elevated, not left dependent A week of this routine should be taken to accustom the leg to new conditions, after which the bandage may gradually be left off

Should an ulcer have been excised and a skin graft made, some weeks are required to accustom the graft to its depend

ent position. It tends at first to be very cyanotic and to break down at its edges. It must therefore be supported by a soft pad and a semi-elastic cotton bandage.

Rupture of Varicose Veins.—Occasionally and unexpectedly, a varicose vein ruptures externally, giving rise to a gush of dark blood. The vein which ruptures does not seem to be the great sacculated vessel projecting above the surface of the leg —such is more apt to undergo thrombosis—but a smaller and less conspicuous one, the wall of which, however, is covered only by very thin skin. Rupture always occurs below the knee. Naturally, for so long as the leg remains dependent, blood flows rapidly from the tiny opening and will in time, if unchecked, drain the entire vascular system. However, the leak is promptly stopped by elevating the leg above the body and making pressure upon the region of the hole. A firm bandage over a small gauze pad sufficiently compresses the varicose vessel so that little or no blood can reach the opening. Thus the individual, if given first-aid treatment, can usually get about at once. It is then advisable, not as an emergency but within a week or two, to divide the vein above the rupture, whether or *not the* great saphenous is resected at the groin. Rupture is of course an indication for some sort of curative treatment of varix.

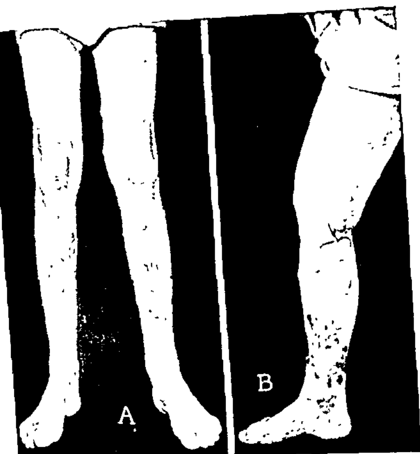
Thrombophlebitis in Varicose Veins.—Those who have read, up to this point, the sections devoted to varix will have observed that almost no mention of thrombosis has been made. The apparent *omission* is due to the writer's conviction that all forms of thrombophlebitis should be considered together —that which occurs in varicose veins being the most common, least dangerous, and on the whole the most tractable form. The thrombophlebitis of varix will, therefore, be described in the following chapter. It is enough to say of it here that it is, next to ulcer, the most annoying and disabling complication of varicose veins. It occurs unpredictably in persons of all sorts, young, old, vigorous and feeble. Once it has attacked a varicose vein it is apt *to return*, like the tiger with a taste for human blood. Its treatment, because it is an everyday affair

which almost never takes life, has long been unenterprising and dictated by outworn traditions Embolism is exceedingly rare but does occur

Postphlebitic Induration and Ulceration

Little is to be found in textbooks or in the literature of ulcer upon the subject of the common disabling indurations and ulcers which follow phlegmasia alba dolens (femoro iliac thrombophlebitis) Leriche briefly describes and pictures them as results of a vasomotor disorder associated with this sort of thrombophlebitis In this country, Trout has noticed them and described an operative treatment The writer, in early papers (1916–17) primarily devoted to varicose veins and ulcer, de scribed them as "the sort of ulceration which is associated with postphlebitic varix of the small vessel type" That is, he was unable to shake off the idea that some sort of varicosity was behind the ulcer, though he recognized the postphlebitic character of the disease Actually this was a useful concep tion, for it led to many efforts to cure the indurations and ulcers by excision of the dilated superficial veins and division of the communicating veins with which the lesions are so often associated But the dissections proved that though de fects of venous drainage were a contributing factor, the lesions could and often did occur in their absence and were not in fact "varicose ulcers" This is the reason for the considerable space given here to these lesions Many practitioners, upon seeing something which appears to be a varicose ulcer, seek out any vein in the vicinity and inject into it a sclerosing solu tion By so doing they merely increase the local inflammation and aggravate the sore

Postphlebitic induration and ulcer follow only a deep throm bophlebitis No enlarged visible veins need be associated with them However, in a minor proportion of cases a number of superficial veins seem to become enlarged, as collaterals, dur ing the obstruction of the deep vessels and afterwards become varicose Moreover, there may rarely be associated with them a straight, hard, shrunken saphenous vein which has apparently



"VARICOSE ECZEMA" *A* L. S., 59957, a man, aged forty. Bilateral varices of single vein type. The "eczematous" area is deep red, moist, scaling. Thought to be an allergic reaction to fungous infection. *B* M. C., 60150, a woman, aged sixty. Varicose veins of large type and a red, weeping area—allergic? The ink mark below the knee indicates the proposed site for low division.

PLATE VI

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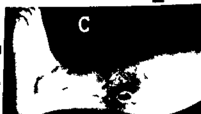


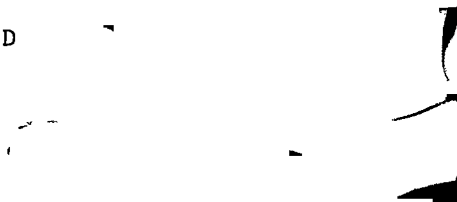
POSTPHLEBITIC INDURATION AND ULCERATION. J. C. H., 50374, a woman, aged forty-nine. A small but deep lesion in a common situation. Shortening of tendo-Achillis. Before operation. *B.* After saphenous nerve division just below knee.

PLATE VIII









POSTPHLEBITIC INDURATION AND ULCERATION. *A*, D. G., 30728, a woman aged forty-two. Secondary varix with unusual degree of sclerosis. Cure by excision of vein. *B*, M. A. H., 3132, a woman, aged sixty-five. Notice crease over sclerosed great saphenous vein secondarily involved in femoro-iliac thrombophlebitis. *C*, M. C. O. D., 23076, a woman aged twenty-three. Pigmentation and ulcer in an unusual situation. Successfully treated by excision and skin graft. *D*, H. K., 59506, a woman, aged forty-seven. Early stage of induration without ulceration. No veins visible. Elevation of flap showed no incompetent communicating veins. Treated by lumbar sympathectomy.

PLATE VII

been thrombosed at the time of the femoral thrombophlebitis. And there are very likely to be associated with them incompetent communicating veins (whether or not dilated superficial veins are evident). Any or all of these may contribute to the persistence of postphlebitic induration and ulceration. However, as has already been explained, it is easy enough to find cases in which obstinate postphlebitic lesions are associated with no abnormal superficial or communicating veins whatever.

Onset and Course.—Because the pathological features and appearance of the lesion itself are not entirely characteristic, its onset and course, which *are* peculiar, will first be described. The earliest signs of the disease appear at any moment from three months to twenty years following the femoroiliac thrombophlebitis. The "milk-leg", "phlebitis", or whatever it may have been called, will usually have been severe, but recovery will not necessarily have been, though actually it often is, succeeded by residual swelling. Without warning and usually with little discomfort, a patch of edema will be noticed upon the lower third of the leg, most often upon the inner face of the calf a few inches above the ankle, but occasionally upon the back or in the neighborhood of either malleolus. As a rule, the swelling is low, rounded, hot to the touch, very slightly reddened, and from two to four inches in diameter. One seldom sees the lesion at this early stage, since the patient does not consider it serious. Actually it is nothing but a local patch of edema, not a furuncle-like affair, yet it is slightly hot to the touch as if it were the seat of a mild, nonsuppurative infection. In some instances, the appearance of swelling is lacking and the skin is merely pigmented and faintly indurated. If pigmentation is the first sign, the color may deepen rapidly to a heavy brown. Rarely pigmented patches or indurated areas are multiple.

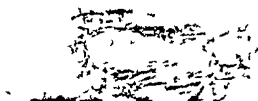
If seen very early, the local edema can usually be made to disappear upon elevation for a few days in bed. The writer has seen one or two cases aborted, as it were, by prompt elevation and bandaging. The following is an instance of this sort:





A

B



Postphlebitic Induration and Ulceration. I. S., 30633, a woman aged fifty-three. *A*. Small ulcer but larger area of pigmentation and induration. Note small dark areas toward back of calf separate from the main lesion—each a local induration of fat. *B*. Two years after excision and Ollier-Thiersch graft.

Except for its relation to phlegmasia alba dolens and its mode of onset, that is, through the establishment of local edema and induration in the absence of varicose veins, there is little to distinguish it from any other ulcerative process upon the legs. Pain is usually present, especially in the case of lesions upon the inner face of the calf, a little above the internal malleolus. Such ulcers have been called "irritable ulcers" and are often agonizingly painful.

Veins are rarely noticeable, at least at first, and, indeed, may never appear. However, as already explained, an occasional collateral enlargement may have ended in varicosity, so that a rather sclerosed type of varicose vein sometimes leads from the groin to the region of the induration and ulcer. In that case, varicosity clearly contributes to the development of the sore and gives an opportunity to test for the presence of incompetent communicating veins. These latter will often be found to be present, so that it has seemed to some as if they must be a basic feature of the disease. Their presence is discovered by using the second form of the Trendelenburg test, that is, the test of back-flow when a constriction is applied to the thigh. On lowering the leg, after a turn of bandage has been tightened about the thigh, the veins of the calf will be felt to become tense in five to ten seconds, or even less, showing that venous blood is leaking from the deep veins out through the communicating vessels to the surface. But the foot does not become blue; for the deep veins themselves are not crippled.

Undoubtedly the leaking communicating veins are frequently a feature of postphlebitic ulcer, and one or more will be found (on exploration) beneath a great area of induration. However, the writer has excised many such areas in toto, including the aponeurosis beneath, without finding any perforating veins of an incompetent sort (one can test their competency at the operating table by observing whether or not they are grossly dilated and allow blood to flow toward the surface when cut). Indeed it has never seemed to him that the veins were greatly concerned with the pathology of the lesion.

A married woman, thirty three years of age, had suffered, seven years earlier, from a bilateral milk leg, the left by far the more serious She had subsequently gone through a pelvic operation without a recurrence, but the left ankle had always swollen after a long day on her feet For several days before she came under observation, she had noticed a localized swelling and hardness just above the external malleolus This gave her some discomfort, especially at the end of the day In other respects she was well

Examination revealed an area of indurated edema several cm in diameter, slightly reddened and barely elevated above the surrounding skin which, as compared with the other foot, was faintly pinkish blue in color

A semi elastic cotton bandage was applied from toes to knee and the patient was advised to spend as much as possible of the next week in bed This she did, with the result that the ankle took on a natural color and the edematous spot disappeared She was instructed that the early lesion was a danger signal and that if at any time she expected to be much on her feet she should use her bandage Two years later she reported that she had had no further trouble, barring some swelling of the ankle when her children kept her particularly busy Whether early treatment should always produce this favorable result is unknown, for most patients present themselves only when the edema has become permanent induration or actual ulceration

Once the lesion is established, it takes on a discolored appearance, usually a combination of pigmentation and redness which fades into the normal tissues about it The skin is thickened, the subcutaneous tissues hardened. The hardness fades gradually, in some cases, into the normal soft quality of the subcutaneous fat In others, it ends irregularly and abruptly so that its rather scalloped border, though invisible, can be palpated At this stage, ulceration will usually have occurred at its center, a sore very much like the early varicose ulcer, having its same tendency to heal on elevation and protection, and the same tendency to widen and deepen with time

calf. On the other hand, the very early lesion, as in the case already quoted, may be made by elevation, rest and bandaging to disappear altogether, or, if more advanced, will by similar methods be held in check. In some instances, enlarged veins, passing into the indurated area from above, can be removed with benefit, an operation which may include resection of the entire great saphenous system. At the same time any incompetent communicating veins encountered can be divided at the level of the aponeurosis. Any such operation, it will be realized, will approach, if it does not actually enter, a field in which the tissues are indurated and perhaps actually infected. It is carried out exactly like the operation for varicose ulcer (*q.v.*). It will be best that any such operation should not be performed in the presence of an open ulcer—the tissues are already sufficiently liable to infection. They must be handled with great gentleness, and asepsis must be perfect. Recently, Linton has shown how by long incisions from knee to ankle, carried through the aponeurosis, the area of induration can be elevated and the incompetent perforating veins divided from beneath.

Excision of the indurated, ulcerated area is a very satisfactory way of permanently curing the disease. This does away at one moment with any venous stasis and the badly diseased tissues. The operation should be reserved for cases with heavy and wide-spread induration, should remove *all* scarred tissue, no matter how large the area, and should always include the muscular aponeurosis. If this heavily scarred layer is not taken, a skin graft upon its surface will never permanently survive. On the other hand, if the aponeurosis is removed, a successful graft can be placed upon the exposed periosteum, tendon-sheath, muscle, or capsule of the ankle joint—the tissues beneath the aponeurosis not being involved. The excision should be performed in a bloodless field (Esmarch bandage to thigh). After removal of the Esmarch bandage, the bleeding vessels should be tied with the finest silk. An Ollier-Thiersch graft, immediately applied, will almost invariably "take" and survive, though its subsequent adjustment to a dependent posi-

The most noticeable *pathological feature* of postphlebitic ulceration is the thickening and hardening of the tissues, which is most marked upon the surface of the aponeurosis Indeed, this layer is sometimes two to four mm in thickness, a dense leathery barrier through which the arterial supply fails to penetrate from beneath (the superficial tissues are supplied, as already explained, by arterial "trees" which pass from the great arteries out through the aponeurosis in a series of rows) If, then, one were to sketch what appear to be the steps which lead to the establishment of the advanced lesion, one would place them in the following order localized edema (lymphatic or vasomotor?) associated with low grade infection, fibrosis, lowered resistance to infection, ulceration After which the vicious circle of edema, fibrosis, diminished arterial supply, and infection continue and cause the lesion to extend Sometimes, indeed often, after a great ulcer has been excised and skin-grafted, yet leaving a little induration at one edge, an acute process will recur at that point and soon involve new areas

The diagnosis is suggested by the appearance of the lesion and the absence of a history of varicose veins Indurations and ulcers unusually placed (from the standpoint of varicosity) or multiple are nearly certain to be postphlebitic The accompanying page of illustrations will support the statement that the appearance of any one lesion should lead to an investigation of its background See Plates VII, VIII and IX)

Treatment—Since induration and ulceration may become established in youth, following the milk leg of childbirth and the thrombophlebitis of acute fevers, surgical operations and injuries, there is often granted an opportunity for cure at a time when the tissues are still capable of permanent healing Individuals first seen when over fifty years of age, especially when the lesions themselves have been present for many years, are usually incurable Indeed, there is nothing more resistant to treatment than a long standing postphlebitic ulcer which occupies the inner face of the leg from ankle half way to knee and encroaches on both the anterior and posterior faces of the

pathectomy, had better be treated by other methods. In the female, removal of the first lumbar ganglion is not only harmless but will raise the level of vasodilatation to the thigh.

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tion is often time consuming It is remarkable how soon the abrupt edge left by the excision of such an area is flattened and smoothed, showing that the edema and thickening about the ulcerated area has disappeared

Other, less radical means of dealing with these postphlebitic lesions are (1) nerve division and (2) lumbar sympathectomy The former is only available for painful, but not too deeply scarred processes upon the inner face of the calf, at or above the median malleolus The latter is most likely to succeed in the case of lesions moderately indurated

Internal Saphenous Nerve Division is especially useful in treating painful ulcers in the field of that nerve The ulcer should be healed or at least very clean when the operation is performed A transverse incision is made under procaine infiltration just below the crease on the inner face of the knee joint, that is, over the great saphenous vein which exactly overlies the nerve and is the guide to it The nerve, in turn, lies upon the aponeurosis, exactly behind the vein, usually a single structure but sometimes in the form of two trunks (having split just above) Having divided the nerve, the operator will do well to bury its proximal stump beneath the aponeurosis On the whole, the larger the nerve and the less it has broken up, the more clean cut and free from overlapping collateral sensory supply will be the saphenous field

The effect of a saphenous block is to raise slightly the cutaneous temperature (partial sympathetic paralysis) in the saphenous field The ulcer will become painless and will very often heal (See Plate VIII)

Lumbar Sympathectomy is only likely to succeed when induration is of moderate grade, incompetent superficial and communicating veins are absent, and the vasodilatation of the sympathetic paralysis can be made to mount higher than the lesion To cause vasoparalysis to reach up to the knee, excision of the first as well as the second and third lumbar ganglions will usually be required, an operation which may disturb the male sexual function Therefore, in the male, indurations which extend high, though otherwise appropriate for sym

pathectomy, had better be treated by other methods. In the female, removal of the first lumbar ganglion is not only harmless but will raise the level of vasodilatation to the thigh.

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CHAPTER VI

THROMBOPHLEBITIS AND PULMONARY EMBOLISM

The thrombophlebitis of surgical operation, accident and serious illness is recognized today as a problem of the first importance. This venous thrombosis is one that occurs, not in the auricles of the heart, the cerebral sinuses, or the portal system, but in the veins of the pelvis and lower limbs, a sort which attacks persons put to bed because of surgical operation, childbirth, injury, or disabling disease and which not only adds a new illness, but introduces the hazard due to the presence of a detachable thrombus. Beside this common variety, which might well be called the thrombophlebitis of hospitalization, there are other forms which occur in active life. Each has its anatomic background and each presents itself in a characteristic way.

The word, thrombophlebitis, is taken to mean thrombosis in a vein and does not necessarily imply inflammation, certainly not such [illegible] tion. "Phlebitis," s [illegible] word, thrombosis being the basis of the process and phlebitis a reference to its scene. At its very start, thrombosis is not clotting; for clotting relates *only* to coagulation of the blood, a chemical-biological process which follows fairly well-defined rules, occurring when blood is *exposed* to tissue juices, after death or in association with the release of a thromboplastic substance from thrombocytes. Thrombosis can and usually does take place within normal intact blood vessels. The thrombocyte, or platelet, is the villain of the piece, attaching itself in places where its death will be *most* inconvenient. When a mass of these tiny discs becomes plastered *upon the* intima of a vein, they die, and a thrombus has begun to form.

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unnecessary to follow what has become the classical form, and relate the almost innumerable causes of thrombosis assembled under Lubarsch's tripos, that is, disorders of the venous return, disorders of the blood, and disorders of the vein's wall. An account will be given rather of the factors which are recognized today as leading most directly to thrombosis and which are being attacked with some vigor in various parts of the world. These include: (1) retardation of the venous flow, and with this certain basic anatomical relations; (2) dehydrating factors; (3) the local and general influence of trauma, which may be called the "X" factor; and (4) inflammatory changes about certain great arteries and veins.

In the discussion of thrombophilic influences which follows, reference will necessarily be made to several varieties of thrombophlebitis. These will subsequently be fully described. It is sufficient here to give a brief characterization of each.

Femoro-iliac Thrombophlebitis: Phlegmasia Alba Dolens: Milk Leg.—This is the common "phlebitis" which is responsible for the great, white, swollen leg—usually an outspoken disease, sometimes painful, especially at its onset. Its scene is the principal vein draining the leg in the region of the groin. Only a very small proportion of femoro-iliac thromboses lead to pulmonary infarction or serious embolism. Yet the disease is so common that the number of resulting embolisms must be considerable.

Thrombophlebitis in the Deep Veins of the Lower Leg.—This seldom-recognized disease affects the great plexuses of veins in and among the muscles of the calf*—a system required for emergency use and therefore capable of nourishing, with little external sign of congestion, a silent, treacherous process—a frequent cause of embolism.

Thrombophlebitis in Nonvaricose Superficial Veins, a freakish process, due to trivial causes, often local but an occasional cause of a propagating thrombus and at least minor pulmonary embolism.

* A good idea of the capacity of these veins can be had by consulting Figure 13 (Chapter V) and Figure 220 of this chapter.

The thrombus grows into the venous stream as a sponge like mass of dead platelets, which excites coagulation, so that leucocytes, red cells and fibrin are soon entangled in its tough meshes At first the process does not close the vein but when the body of the thrombus has been built out from the adherent head, the vessel is soon filled with a mixed, dark red solid mass As the thrombus extends up and down the vein, its youngest portion, or tail, is seen to be soft and clot like having no longer a supporting framework of platelets Indeed, the tail of a thrombus is almost pure coagulum, red and flimsy, easily broken up and carried away A thrombus tends to grow until it meets a vigorous stream, and thus its proximal end is likely to heal at the point where a branch carrying a strong current enters the thrombosed vessel Its extension distally is uncertain, since it is not easy to say where a good current will be able to leave the main vessel against the set of the valves in the entering branches However, there are many curious bypaths in the venous system, and doubtless the establishment of collateral channels is less difficult than one would suppose In a complete backwater, the intima being intact, a thrombus does not form It is the slow and feeble current which offers it encouragement Into such a sluggish stream, proximal to the main thrombus, the flimsy tail grows on, often projecting from a smaller vein into a greater or along a greater in the slow, entering current of a smaller—a propagating clot

This propagating clot, soft and fragile, waving free in a large vein, such as the femoral or external iliac, is the source of pulmonary embolism, for the embolus, once broken off, meets with no obstacle from its point of detachment in its course through the great iliac vessel, the vena cava, the right side of the heart, and so into the pulmonary artery

It is easy enough to grasp the nature of the thrombus, its solid head, occluding mixed body and insecure tail But why do thrombocytes lay themselves down to die in particular parts of the venous tree? And why, under circumstances equally favorable to thrombosis, does a thrombus form in one individual and not in fifty more? In presenting this problem, it seems

confused, encouraging the settling and adherence of thrombocytes. Another area, more commonly the scene of thrombophlebitis than is generally supposed, is the popliteal region and the upper part of the calf. For several plexuses draining the great, flat, flexor muscles come together here.

Most, if not all of the causes of a slow venous return disappear when the legs are elevated, especially if the thighs are not flexed but in line with the body. For this reason, an obvious first step in thrombus-prevention is elevation of the foot of the patient's bed. Already some observations have been made upon the favorable effect of such treatment, especially in gynecologic surgery, and since many surgeons are attempting by this method to discourage postoperative thrombosis, positive information of its value will soon doubtless be available. There is also evidence that a thrombosis once started ceases to progress toward the heart when once it meets a vigorous current. For example, in the varicose saphenous vein it frequently ascends to the junction of that vein with the femoral, which, however, it is almost never able to enter. The writer has observed, moreover, on several occasions, the effect of elevation upon superficial thrombosis in a nonvaricose vein. In an individual whose thrombosing process had just run over both legs while he was allowed to recline in bed, a new thrombosis was halted and disappeared within a week when the foot of the bed was elevated six inches.

By contrast, as already explained, the dangerous propagating thrombus, the source of fatal pulmonary embolism, is fostered by a slow current. That is to say, the friable, clot-like tail of the thrombus grows out into a feeble stream entering from a proximal branch or, if thrombosis has begun in a branch, the tail grows into and waves in, without occluding, the sluggish current of the principal vein. Until the propagating thrombus has been studied further, the conditions under which it forms and grows can only be surmised, yet there is evidence, especially in instances of thrombosis in the deep veins of the calf, bearing upon the matter, as will appear in the following accounts.

Thrombophlebitis in Varicose Veins, a common disease, incapacitating but not dangerous. The thrombosis is usually solid and strictly confined to the varicose vein—rarely a source of embolism.

Thrombosis also occurs in the venous plexuses of the internal genitals of both sexes, the prostate and the uterus, but the behavior of the process is little understood and its clinical nature is unknown. It is probably a source of serious pulmonary embolism.

Retardation of the Venous Return, that is, a slow but not a dead current, is essential to thrombosis. In surgery, the retardation is usually due to mechanical causes, in medicine, to debilitating disease or enfeeblement of the heart. But in any case, confinement to bed is of first importance. Certain anatomical relations are of hardly less moment. Blood is pushed out of the lower limbs by muscular action. Therefore, a patient reclining or sitting up in bed causes the return flow from the relaxed legs to be decidedly delayed. Add to this effect the increased abdominal tension of intestinal distension, post operative or otherwise, of tight abdominal dressings, and of excessive adiposity. Or add to it the pelvic venous congestion of pregnancy and the puerperium. Such influences aggravate the inherent difficulty of emptying the veins of the legs and pelvis unless the lower part of the body is raised above the upper.

Certain anatomical features not only retard the venous blood flow but introduce those eddies and cross currents which, according to many, favor a deposit of blood platelets in certain localities. The relation of the iliac veins to the great arteries of the pelvis is sufficiently familiar. The left common iliac vein is crossed at almost a right angle by the right common iliac artery. There is thus a hypothetical slowing of the venous current in the left common iliac vein. The slowing is perhaps most likely to produce its effect where the vein passes behind the hypogastric artery. In the region of the groin, many branches enter the femoral and external iliac veins. Such valves as may be present are large. Here, then, the current is

which are not obstructed by the initial thrombosis. It is very probable, though by no means certain, that the same result is to be expected in a majority of similar cases.

To sum up the disorders of the venous return: the difficulty of forwarding blood from the legs is increased by a reclining position in bed, especially if the legs are flaccid; it is aggravated by increased abdominal tension and pelvic congestion; and because of confused currents in the upper calf and at the groin, a slow stream is especially likely to lead to thrombosis at these points; finally, a slow current, once thrombosis has taken place, favors the formation of a dangerous propagating clot, just as a brisk current discourages such a process.

Dehydration has long been recognized as leading to thrombosis. Individuals who have become anemic because of bleeding uterine fibroids, those who have become depleted by vomiting, by sweating and failure of fluid intake in connection with an abdominal operation, perhaps those also who have suffered serious malnutrition from any cause, are more liable than others to thrombophlebitis. Dehydration must, however, be counted among the influences which occasion clotting rather than thrombosis, in the strict sense of the words, influences which Bancroft, of New York, has described so well and for which he has established a "clotting index". On the whole, dehydration and depletion of the blood in general are the least obscure and most remediable of all the various causes of thrombosis under discussion. Means of prevention, of course, can readily be practised. The free administration of parenteral fluids, if the intestinal tract is not sufficiently available, and, when necessary, transfusion of blood, offer the obvious remedy. Further discussion of thrombophilic influences in the blood and body fluids will be found in the following section.

Trauma, the "X" factor, which seems to be the immediate excitant of thrombosis, is not only among the most active but certainly the most elusive of all thrombophilic influences. Its importance today is attested by the fact that such measures as are directed against thrombophlebitis are being used immediately after the operation, childbirth, or accident. Though

A man, fifty years of age, suffered a fracture of the fifth metatarsal bone in a minor accident A plaster cast was applied for a week For the next four months swelling of the ankle and moderate discomfort in the calf occurred repeatedly upon use of the leg and rapidly disappeared upon rest in bed At the end of this period he died of pulmonary embolism The twelve to fifteen inch (thirty to thirty five cm) embolus was found to have been detached from the point at which a large thrombosed vein entered the popliteal, as shown in Figure 22C The femoral vein had never been occluded Here was a fatal easily detached, propagating clot, encouraged to form by partial occlusion of, and retardation of the current in, the venous tree and never subjected to continuous elevation By contrast, prompt elevation following the establishment of thrombosis in these same vessels seems to have a favorable effect and is likely to cause the thrombosing process to recede and heal The following is a case in point

A vigorous athletic man, twenty seven years of age, was first seen when, some four days after jumping a brook, one leg had become lame, the ankle and calf considerably swollen but without any ecchymosis He displayed the sign which the writer believes to be characteristic of this disease, *that is*, a painful soreness noticed high up in the back of the calf upon forced dorsiflexion of the foot, by which the tendo Achillis is put on the stretch The foot of his bed was elevated six inches, the leg placed on a soft pillow but not immobilized. In a week, the swelling had disappeared In ten days, all soreness on passive dorsiflexion of the foot had gone, and in three weeks the patient was going about as usual There has been no recurrence

This, like the preceding case, is judged to represent thrombosis in one or more of the great intermuscular plexuses of the lower leg But here, instead of resulting in the formation of a propagating clot and death from pulmonary embolism, the thrombosing process healed to be organized and absorbed This favorable outcome is laid to elevation and the establishment of a brisk venous current in the many veins of the region

and fibula. The suggestion has been offered, without proof, of course, that some product—allergic or otherwise—of damage to, or rapid atrophy of, muscle, consequent upon the injury, is the active, exciting thrombosing influence.

There is then, perhaps, a state of the blood, related to the general and local effect of trauma, which may be expected to act rather rapidly. The conclusion is difficult to escape that when other influences are favorable, the injury determines the decisive action of the thrombocyte. Best and his associates in Toronto have attacked thrombocytic adherence directly, making use of a perfectly pure heparin, which they have developed themselves. By intravenous therapy, based upon animal experimentation, they have a reasonable hope of preventing the adhesion of the thrombocyte to a venous surface. Sufficient heparin to raise the clotting time to about fifteen minutes has been given continuously for several days, beginning an hour or so after operation, in a long series of major procedures. Apparently, postoperative hemorrhage is not a danger and thrombosis is abolished. Similar observations, which have not as yet been sufficiently prolonged to lead to an authoritative conclusion, are being made by Crafoord in Stockholm. One may say, then, that hypothetical changes in the blood, related to trauma, are important enough to suggest the need of using measures directed against thrombosis immediately after the operation or other injury. This, of course, is not at all new. Previous attempts, such as the administration of desiccated thyroid gland, exercises in bed, and, more recently in Europe, "sympatol" and other substances have been used in a similar way.

Perivenous Inflammation.—Changes in the vein's wall, brought forward as one of the causes of thrombophlebitis, have been thought to represent degenerative and inflammatory reactions. In the presence of acute fevers, this seems intelligible enough, and indeed acute rheumatic changes, actually leading to thrombosis, are recognizable. However, evidence of such lesions in the vein's wall as a consistent cause of the familiar, and especially the postoperative, forms of throm-

the gross evidence of thrombosis, that is, edema in the form of phlegmasia alba dolens, may not appear until two to three weeks or even longer after operation, there is good reason to believe that the actual onset of thrombosis and of the conditions which lead to it occur much earlier Moreover, if the propagating thrombus must, as Aschoff states, attain a length of some twelve inches (thirty cm) or more to become a fatal pulmonary embolus, it clearly requires time * for its formation, presupposing an onset of thrombosis very early indeed Thus, although the proper combination of factors can surely occasion thrombosis at any time, the early hours and days are especially liable to it

The thrombophilic influence of trauma is, apparently, both general and local There is no intelligible reason why an operation upon the gall bladder should cause a thrombosis in the left upper femoral and external iliac vein (its favorite seat) Yet unless the writer has been completely deceived, he has seen a locally reactionless thrombosis start in such a patient on the night of an utterly uncomplicated cholecystectomy and lead to a fatal embolism three days later (as proved by autopsy) Here the very fact of operation, barring, perhaps, increased abdominal tension, seems to have been the only exciting factor But it is well known, also, that thrombophlebitis following fractures of the bones of the thigh and leg almost always occurs on the injured, not the sound, side This is brought out in Vance's account of the fatal embolisms of accident cases observed over a period of ten years by the Chief Medical Examiner's Office in New York City He reports that, in the large majority of instances, the causal thrombosis was found on the side of injury, however slight that might be But one does not need to assume that actual damage to the vein itself is responsible, since fractures of the femoral neck are nearly as apt to lead to thrombosis as are those of the tibia

* Fatal embolisms have been known to occur on the evening of the day of operation. They are by no means rare on the third or fourth day Therefore a long propagating thrombus must be able under the right conditions to form with great rapidity

and fibula. The suggestion has been offered, without proof, of course, that some product—allergic or otherwise—of damage to, or rapid atrophy of, muscle, consequent upon the injury, is the active, exciting thrombosing influence.

There is then, perhaps, a state of the blood, related to the general and local effect of trauma, which may be expected to act rather rapidly. The conclusion is difficult to escape that when other influences are favorable, the injury determines the decisive action of the thrombocyte. Best and his associates in Toronto have attacked thrombocytic adherence directly, making use of a perfectly pure heparin, which they have developed themselves. By intravenous therapy, based upon animal experimentation, they have a reasonable hope of preventing the adhesion of the thrombocyte to a venous surface. Sufficient heparin to raise the clotting time to about fifteen minutes has been given continuously for several days, beginning an hour or so after operation, in a long series of major procedures. Apparently, postoperative hemorrhage is not a danger and thrombosis is abolished. Similar observations, which have not as yet been sufficiently prolonged to lead to an authoritative conclusion, are being made by Crafoord in Stockholm. One may say, then, that hypothetical changes in the blood, related to trauma, are important enough to suggest the need of using measures directed against thrombosis immediately after the operation or other injury. This, of course, is not at all new. Previous attempts, such as the administration of desiccated thyroid gland, exercises in bed, and, more recently in Europe, "sympatol" and other substances have been used in a similar way.

Perivenous Inflammation.—Changes in the vein's wall, brought forward as one of the causes of thrombophlebitis, have been thought to represent degenerative and inflammatory reactions. In the presence of acute fevers, this seems intelligible enough, and indeed acute rheumatic changes, actually leading to thrombosis, are recognizable. However, evidence of such lesions in the vein's wall as a consistent cause of the familiar, and especially the postoperative, forms of throm-

bophlebitis is not forthcoming Moreover, there seems to be no difference, either clinically or pathologically, between the thrombophlebitis complicating acute fevers and that associated with traumatic, postoperative and puerperal states Evidently infection and fever, as related to the operation or injury it self, are not important etiological factors in thrombophlebitis Nevertheless there is a relation between *perivascular* inflammation and thrombosis, a relation unfortunately not often discernible but of such a nature as to make it clear that the inflammation, when present, is a cause of thrombosis and that thrombosis does not occasion the perivascular inflammation

Perivenous inflammation has been discussed by Leriche, not so much as a cause of thrombosis as of the pains, edemas, and late complications attendant upon femoro iliac thrombophlebitis Just as involvement of vasomotor nerve fibers by inflammation of the wall of a large artery leads, he believes, to disorders of the peripheral arterial circulation, so inflammation about a great vein, by involvement of perivenous nerves, causes peripheral pain, edema, and eventually, as a late complication in some cases, ulceration Leriche comes to this conclusion partly upon experimental grounds but chiefly because of noticing the favorable effect of resecting, or perhaps merely freeing, adherent, chronically thickened, occluded iliac veins upon peripheral pain and edema The writer is able to present a somewhat different sort of evidence Being interested in the relation of the lymphatics to the edema of phlegmasia alba dolens, he has, in several instances of this disease, explored the pelvis in order to examine the lymphatic bearing tissues surrounding the great iliac vessels In one instance of a left sided phlegmasia alba dolens, which had already persisted unchanged for about ten days, he found, upon dividing the peritoneum over the iliac vessels, an intense, nonsuppurative, vascular exudate surrounding both artery and vein The artery was so engulfed in inflammatory tissue that it could not be seen, but it was evidently in a state of spasm The vein was only found by cutting into it Strangely enough, its intima was, by contrast with the adventitia, entirely normal and it

was filled with a solid, dark, mixed thrombus. It was clear that a primary perivascular inflammation had led at one moment to venous thrombosis and arterial spasm, and inconceivable that the reactionless thrombus could be the cause of the active, external, inflammatory exudate. The perivascular reaction surrounded the common as well as the left external iliac vessels and was found to extend down Hunter's canal. In the lower third of the thigh it was much less active, and here the femoral vein was found to contain liquid blood. In another case of longer duration, a similar reaction was seen during the stage of repair, the exudate having undergone such fibrosis as to unite the artery and vein firmly to each other and to the surrounding sheath. Yet it cannot be shown that the perivascular reaction is always present; for in a third exploration by the writer, there was no trace of it, though a solid thrombus occupied the left external iliac vein up to the crossing of the hypogastric artery. Obviously, if such a reaction is the rule, it will be necessary to offer an explanation of its origin. One may, therefore, go back to the lymphatics, the primary object of the writer's explorations. Any infection carried by the lymph stream from the legs, the genitals, or the anal region must pass to the lymphatic vessels and nodes about the great iliac blood vessels. And there are many observed instances of lymphangitis associated with the onset of phlegmasia alba dolens. One such case is cited:

A youngish man, suffering from pneumonia of no great severity, exhibited a lymphangitis and femoral adenitis following hypodermoclysis of saline solution in the right thigh. The severe pain in the right leg, which then suddenly set in, was associated with disappearance of the pulses in the right foot and enfeeblement of the femoral pulse. Only after these signs of arterial spasm had occurred did the typical swelling of a femoro-iliac thrombophlebitis appear. Unfortunately this patient could not be followed long enough to determine the further course of the phlegmasia alba dolens.

Here the matter must be left. The more interested one becomes in perivascular inflammation as a cause of femoro-iliac

thrombophlebitis the more important it seems, but the various other causes of thrombosis are clearly so significant that one fears to become too much attached to one—and that little studied—hypothesis However, a quotation from Cruveilhier, upon the subject of iliac thrombophlebitis, is perhaps appropriate

"D'une autre part, j'ai vainement cherché dans la membrane interne des veines des traces d'inflammation point d'injection vasculaire, point d'epaississement notable On ne trouvait d'indices de travail fluxionnaire que dans le tissue cellulaire *exterieur* aux veines, lequel tissue etait plus coherent que de coutume et injecte de capillaires veineux qui enlaçaient la veine comme dans un reseau et penetraient dans son epaisseur Dans aucun cas de phlebite, je n'ai trouve la membrane interne des veines injecte" (Italics, writer's)

Pulmonary Embolism.—Before going on to the specific varieties of thrombophlebitis, it will be appropriate to discuss the nature of pulmonary embolism The *Propagating Thrombus* has already been pictured as the usual source of embolism There is ample reason to believe that, if it is to form, there must be available, proximal to the primary site of the thrombus, a sufficient length of large vein That is, if the solid part of the thrombus ends proximally where the external iliac vein joins the hypogastric, the propagating thrombus must float in the common iliac and far up the vena cava Aschoff believes that most long, fatal emboli come from the femoral, an assumption the more reasonable because of the fact that outspoken phlegmasia alba dolens, which represents a thrombosis extending proximally through or above the external iliac vein, is seldom a source of fatal embolism Certainly, many long emboli are broken off in the popliteal vein, having grown into the femoral as propagating thrombi, whence the importance of thrombosis in the deep veins of the calf

In regard to the pelvic plexuses as sources of emboli, little is actually known Thrombosis has been shown to extend, as an occluding process, from the uterine veins into the hypogastric and common iliac But the unanswered question is Does

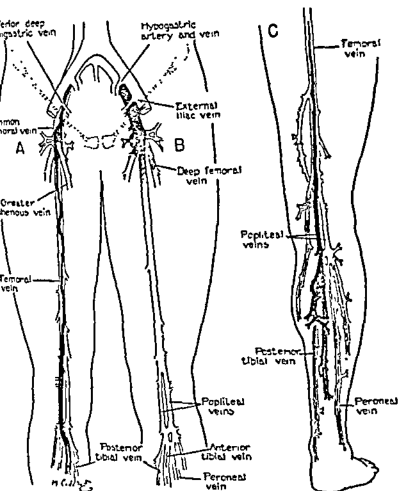


FIGURE 22. THROMBOPHLEBITIS IN THE FEMORO-ILIAC REGION AND DEEP VEINS OF THE LOWER LEG—Semidiagrammatic *A*. A non-obstructing thrombophlebitis such as may be encountered in exploration of the common femoral. It causes little or no swelling and seriously threatens pulmonary embolism. In such a case, the common femoral must be divided and the clot gently sucked out. *B*. Phlegmasia alba dolens, obstructing, and a cause of marked swelling Entering branches more or less involved. Peripheral extent vague. Seldom causes embolism. *C*. Thrombosis of the Deep Veins of the Lower Leg A reconstruction of an actual case in which fatal pulmonary embolism occurred. The long propagating thrombus broke off in the lower femoral. An early division of the superficial femoral vein would have prevented this accident.

a floating, propagating thrombus originate in the uterine or prostatic veins and grow out into the hypogastric and common iliac? If so, it may well break off, leaving little trace behind. Apparently there is no evidence that such occurs, though when no other source of embolism is discovered, the great pelvic plexuses must be under suspicion. Indeed, it is then hard to see from where else the embolus can have come.

Embolism from unnoticed Thrombosis.—There is good reason to believe that outspoken processes such as phlegmasia alba dolens or thrombosis in varicose veins are less apt to cause embolism than the quieter ones. In the case of phlegmasia alba dolens, inflammatory changes, such as have already been described, tend to fix the thrombus, and the collateral current called forth by the obstruction of a considerable length of the main venous trunk would seem to offer little opportunity for the formation of the long, fragile, clot like, detachable tail. Again, in the case of varicose veins, the sclerosed state of the vein, the overstretching of its coats by rapid dilatation, perhaps the presence of bacteria in its wall, combine to make a thrombus adhere solidly. Common as it is for thrombosis, beginning in a sacculation of the upper calf or thigh, to reach the saphenous opening, the extension of a soft propagating clot from the varicose saphenous vein into the femoral is almost unknown. Conversely, among Vance's ninety fatal accident cases of pulmonary embolism, already mentioned, there was rarely any external evidence that thrombosis had occurred. Another pertinent observation relates to the trivial injury as a cause of a quiet thrombosis and a fatal embolism. Vance noticed that among eleven such cases, in six, the thrombosis was on the side of the injury, while in the other five, no source of embolism could be discovered—facts which speak for the ease with which thrombosis can be established by trauma and for the increased frequency of embolism when thrombosis is not extensive enough to have caused external signs, or, for that matter, to have left internal traces.

It seems that not only may a short though dangerous thrombosis remain utterly silent, but even an extensive thrombosis

may be remarkably quiet. The reason for this is not entirely clear but a very gradual spread of the thrombosis is the probable explanation, for the collateral circulation can then keep pace with the obstruction. The writer has seen, in a vigorous elderly man, reclining in bed following prostatectomy, repeated pulmonary infarctions when only a very slight cyanosis of one foot was all that indicated the presence of thrombophlebitis. But when the attempt was made to divide the femoral vein at the groin, thrombosis, *incompletely* obstructing the vessel, was found. A fatal pulmonary embolism followed, and autopsy revealed a bilateral thrombophlebitis which extended proximally, on one side, as high as the common iliac vein. It is certain that incompleteness of occlusion by the thrombus is an important factor in the formation of a propagating clot. The writer has several times met with it in his attempts to divide a vein proximal to the supposed source of embolism. A slow blood-flow past a soft thrombus should certainly offer an ideal soil for the creation of loose detachable fragments.

If the observations of Leriche, Cruveilhier and the writer, in respect to outspoken femoro-iliac thrombophlebitis, are taken seriously, and if the frequency of embolism from quiet or unnoticed thrombosis is at all what has just been suggested, it is fair to suppose that with outspoken phlegmasia alba dolens, an insecure, embolism-threatening thrombus is very unlikely to form, while with quieter, incompletely obstructing thromboses it is always a threat. The explanation is offered that the less the vein's wall is inflamed or injured previous to the onset of, and during the course of, thrombosis, the greater is the probability that an insecure propagating thrombus will be established and that a fatal pulmonary embolus will occur.

Varieties of Thrombophlebitis

Femoro-iliac Thrombophlebitis: Phlegmasia Alba Dolens: Milk Leg.—Though instances of femoro-iliac thrombosis have been observed in persons leading an active life (Barker) the

disease usually attacks those who, for one reason or another, are confined to bed The veins involved are the upper femoral, the external, and perhaps common iliac Apparently the con fused currents due to entering branches and large valves favor thrombosis in this region Aschoff remarks that when both sides are affected, the process is likely to extend, on the left, through the common iliac vein, up to the point where the vein passes under the common iliac artery, but on the right, mounts only to the region of the inguinal ligament The anatomical background for this distinction has already been discussed

Phlegmasia alba dolens is particularly apt to follow opera tion or injury in persons in or beyond middle life But it oc curs so often in young women after childbirth and often enough in young adults of either sex after acute fevers or such a simple operation as appendicectomy, as to make any cate gorical statement about a particular age incidence absurd It would be better to say that a femoro-iliac thrombosis comes to mind when any sort of operation is proposed for an individual of fifty-five years or over After an operation upon the pros tate or hysterectomy for fibroids, it is perhaps twice as fre quent as after upper abdominal procedures Surgeons are apt to forget how very often a phlegmasia alba dolens is asso ciated with debilitating diseases—not only the serious, acute fevers like pneumonia and typhoid but the circulatory failures and advanced organic diseases of any sort Fractures of the lower limbs occasionally bring it on, the injury, plus immo bilization of the leg, plus the reclining position so often used combining to offer an ideal background for the disease Rea sons have already been given for believing that the initial thrombosis usually starts very promptly after favorable con ditions for its establishment have occurred How soon it shows itself thereafter seems to depend upon developments little understood

When a whole lower limb is swollen, it is certain that a femoro iliac thrombophlebitis is present, but absence of edema is no proof that the disease does not exist The amount of edema seems to depend upon the extent of the process, the im

permeability of the plug, the efficiency of the collateral circulation, and the degree to which the deep lymphatics are obstructed. Naturally, a very extensive thrombosis makes the prompt establishment of a collateral circulation a difficult matter, whereas a local one is easily circumvented. An insidious onset and development is consistent with an absence of edema. By contrast, a very acute, active process will cause the limb to swell so tensely, within a period of seventy-two hours, that it will not pit on pressure and is utterly unwieldy. A leg of this sort is not cyanotic and though a sufficiently widespread thrombosis is perfectly capable, by itself, of causing an advanced edema, one cannot help thinking that lymphatic obstruction is very much concerned with the swelling. Paine looks upon an almost pure lymphedema as being exceedingly common. The writer, though understanding the perivascular inflammation which Cruveilhier, Leriche and he have demonstrated in their several ways, to be a probable cause of lymphedema, is aware that a very full swelling of a limb can be present without any perivascular involvement of the lymphatics whatever. This de Takats has experimentally proved.

The onset of phlegmasia alba dolens is more often than not ushered in by pain, sometimes of such severity as to suggest acute arterial ischemia. The pain is referred to the groin, the inner face of the thigh, even to the back of the knee or calf. With this there is apt to be some degree of soreness over the upper femoral region and sometimes above the inguinal ligament. But the swelling may appear insidiously, without pain or at the most with a feeling of numbness or heaviness. As a rule, the pain is preceded for twenty-four hours by a moderate elevation of pulse and a slight rise of temperature. It is not followed by swelling for perhaps another day. Then edema mounts rapidly from the ankle to the groin.

The behavior of the local pulse is of great interest. Allusion has been made earlier to a weakening or disappearance of pulsation in the peripheral vessels and even in the femoral. Evidence of arterial spasm is appearing more and more often in the literature. French observers, in particular, stimulated

perhaps by the writings of Leriche, have described arterio spasm so severe as to cause gangrene of the leg Apparently it is not necessary that a perivascular inflammation should be the cause of this spasm The mere presence of a thrombus in the upper femoral and iliac vein is sufficient A very striking account is that of Gregoire

A woman, forty-nine years of age, had been operated upon for pyosalpinx Three days after operation, swelling and heaviness attacked the left leg but so mildly that the patient did not at first complain On the following morning, however, the leg turned bluish, cold, and numb In an hour, the color had reached the knee and soon the thigh as well was cold, cyanotic and rather marbled, a picture of arterial embolism At the same time, the face became pale, the pulse rapid and the temperature elevated (38 5° C) There was no edema and strangely enough, no pain Cutaneous sensibility was lessened No pulsation could be detected in the femoral distal to a point three cm below the inguinal ligament—the usual findings when the femoral is obstructed at its bifurcation by an embolus Upon exploration, the femoral artery was found to be in a state of violent spasm for a distance of only two cm and contracted to the size of the radial This contraction lay beside the lower end of an enormous, bluish black stretch of femoral vein The thrombosis ended cleanly three cm below the inguinal ligament and disappeared beneath the ligament, above which it was not followed Distal to the thrombus, the vein was small and pale When punctured by a needle, the contracted artery bled in spurts Evidently it transmitted a little blood and an injection of novocaine into its outer coats caused it to expand to its natural diameter, bringing back the peripheral pulse However, the vessel soon contracted to its previous size, nor could it again be made to dilate in spite of a periarterial and perivenous sympathectomy over a distance of ten cm (four inches) Perhaps spasm had now set in higher up In any case, during the following days the leg became gangrenous and eventually was amputated above the knee

It should then be recognized that venous thromboses may be associated with, if not actually a reflex cause of, arterial spasm serious enough in some cases to induce gangrene of the limb, and that pain, which usually marks the onset of arterial ischemia, may occasionally be replaced by mere coldness and numbness.

Minor degrees of this reflex sympathetic disorder are not so uncommon. Leriche has made the very interesting observation that it can be broken up by injections of novocaine into the paravertebral lumbar sympathetic. He states that by daily injections discomfort is diminished and that the edema disappears far more rapidly than the ordinary expectation. Whether such treatment should be made a routine in phlegmasia alba dolens or should only be used when evidence of reflex arterial spasm is observed is not yet clear.

The course of a femoro-iliac thrombophlebitis is extraordinarily variable. A mild form causes only a moderate swelling which disappears after ten days and leads to almost no residual edema when the patient first gets about. A severe form, associated with fever and both local and general discomfort, results in a huge, hard leg which changes little over many weeks or even months. When at last the swelling goes down, the patient is sometimes left with a limb larger than its mate and subject throughout life to some degree of edema, especially of the ankle and lower leg. Once a femoro-iliac thrombosis is established, some individuals seem to become increasingly thrombophilic. The opposite limb is involved far more often than is generally supposed, but perhaps with so little swelling that the second process is overlooked. Occasionally a thrombophlebitis passes back and forth from one leg to another, recurring later in the one first attacked.

The appearance of the leg is usually one of white swelling. There may be a faintly cyanotic, pinkish color as if there were present a [illegible] rarely visi[illegible] collateral [illegible] appear in the upper thigh, the pubic region and the lower abdomen, remaining throughout

life Sometimes the *great saphenous vein can vaguely* be felt as a tender thrombosed cord which later becomes varicose Far more often, tenderness over the upper femoral canal gives rise to the suspicion of a superficial thrombophlebitis which, in fact, does not exist The leg is not noticeably hot or cool Only very occasionally are any lymphatic streaks to be seen

The aftermath of a phlegmasia alba dolens is usually far less disabling than might be supposed It is inconceivable, of course, that any valves involved in thrombosis can again function normally, yet very few feet are left cyanotic after the upright position is resumed The ankle may be puffy but the toes are not blue Only occasionally, large veins are left which in time become varicose The probability is that the worst of the thrombosis takes place in the external iliac, where only one valve is occasionally present, and in the upper femoral where after all only a few are lost Collateral vessels help out and the deep veins of the *lower* leg are seldom involved The real difficulty is with the superficial tissues Whether or not these are left edematous, there may develop later areas of edema, of pigmentation, of redness, of induration, and finally of ulceration, the "postphlebitic induration and ulceration" the nature of which is so obscure and the exact cause of which is so hard to understand Very rarely a painful hypersensitiveness, associated, perhaps, with signs of vasospasm is left, a state reminiscent of causalgia (*q v*)

Treatment, Preventive —Certain influences favoring thrombosis are unavoidable in particular, the anatomic and physiologic peculiarities of the venous return from the legs, the exciting factor of operation, injury, childbirth, or debilitating disease, and of course life in bed To these are added the more or less avoidable influences of dependency and immobilization of the lower limbs, increased abdominal tension and dehydration The first set can be minimized, the second, in most cases, eliminated Elevation of the legs opposes the anatomic and physiologic difficulty with the venous return The legs and indeed the body in general can be exercised, *preventing* relaxation and atrophy Increased intra abdominal tension can

be forestalled by perfect closure of wounds, loose dressings and the skillful anticipation of intestinal distension. The difficulty with all such measures is that in nineteen cases out of twenty they are not required—one must go gunning for the twentieth case! One must never, however, leave a patient weakened by operation or disease sitting up in bed for more than a few hours at a time. A permanent sitting position, the legs being relaxed, is an invitation to thrombosis of the quiet type which so often causes embolism. It would be best that the patient about whom one is especially nervous should remain supine or even head downward until he is ready to begin getting out of bed. Indeed, unless the patient has a very vulnerable cardio-respiratory system, an elderly man subjected to prostatectomy had better be kept, during the post-operative period, head down and feet up for a good part of each twenty-four hours.

The prevention of increased intra-abdominal tension has been dealt with by Bancroft and his associates. They insist that abdominal wounds be so carefully closed that tight strapping and binders are not needed and they regard the prompt restoration of intestinal tone by the early use of semi-solid or solid food as essential. Probably the surgeon's handling of the abdominal viscera at the operating table is equally important. An adequate fluid balance, as Maddock and Coller have proved, merely requires an intelligent calculation. In these days, glucose (five per cent) and physiologic saline solution are given, intravenously for the most part, according to the particular indications.

Measures directed against thrombosis should start at the earliest possible moment, not several days after the exciting factor has appeared. This has been realized by Murray and Best in giving purified heparin after major operations in Toronto. By establishing, within an hour or two of operation, a continuous intravenous injection at an appropriate dosage, they raise the coagulation time to fifteen minutes, maintaining it there for several days, and have decidedly lowered, in a long series of cases, the incidence of thrombosis and em-

bolism The presence of heparin not only delays coagulation but prevents adhesion of the thrombocyte, that is, it completely abolishes thrombosis Unfortunately, the treatment is very expensive, and is certainly not yet available for general use One may perhaps depend upon Bancroft's clotting index as a measure of the threat of thrombosis, taking special precautions or using an anticoagulant drug for the case in which thrombosis seems imminent For an account of the test, Bancroft's writings should be consulted His formula is shown below

$$\frac{\text{Prothrombin (1)} + \text{Fibrinogin (0 5 to 0 7)}}{\text{Antithrombin (1)}} = 0\,5\text{–}0\,7$$

An index of over 1 points to a tendency to thrombosis and the need of preparing the patient by the use of a high protein, low fat and carbohydrate diet Whether or not this diet is given, sodium thiosulphate in ten per cent solution is administered intravenously for three consecutive days in a daily dose of ten ccm

The Treatment of Established Thrombosis must always hurry the return of blood from the legs and pelvis A solid thrombus, it must be supposed, occupies the upper femoral and more or less of the iliac vein But is a loose, detachable, propagating clot present at the proximal end of the thrombus? In the usual outspoken phlegmasia alba dolens it is very rarely present in the occasional quiet, barely noticeable and perhaps incompletely obstructing thrombophlebitis it may well be However, the only proof of its existence is the occurrence of a pulmonary infarction due to a nonfatal embolus Therefore, unless one believes that the common iliac vein or vena cava should be divided rather often on suspicion (!) the presence of the propagating process must be ignored and all efforts directed against its *formation* It isn't the patient's turning over in bed which should be blamed for the fatal embolism, but the presence of the detachable embolus, and

the best practical defense against the formation of the embolus is elevation of the lower limbs.

Elevation of the legs has two objects: the hurrying of a collateral stream past the proximal end of the thrombus, to prevent the growth of the fragile clot which a slow stream encourages, and the relief of edema. There is no reason why the leg should not be elevated and no reason against its being moved, the proximal end of the thrombus being within the pelvis and little influenced by such factors. Indeed, why worry about casual exercise when the patient must practice daily the athletic feat of using the bed pan? So the foot of the bed should be raised six inches and the swollen leg elevated still more upon an inclined plane or in a sling. A couple of pillows may be placed under the shoulders and head but the body should not be bent by raising the upper half of the adjustable bed. Under these conditions, the leg will have freedom of motion and should not be covered with ice bags. Beyond the fact that heat usually brings more comfort—if the thigh is painful—the ice bag delays the flow of blood rather than hastens it and occasionally inflicts a frost-bite. The old custom was to sit the patient up in bed, apply ice to the flaccid legs and wait for swelling and fever to subside. Whether the existence of fever is actually an important consideration is unknown to the writer. When present, it may be expected to disappear with the edema. Under the system here described, as edema disappears, active exercise of the leg or legs in bed is begun and should be continued, with assistance if necessary, for some days before the patient is encouraged to get up. Then, with bandages applied up to the knees, he begins to walk, going to bed between his early attempts. From this time on, the use of the legs must slowly and regularly be increased. A return of edema is of course a sign of too much dependence and too little elevation. Standing, or sitting with the legs dependent, encourages swelling. Muscular exercise diminishes it.

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Elevation of the legs has two objects: the hurrying of a collateral stream past the proximal end of the thrombus, to prevent the growth of the fragile clot which a slow stream encourages, and the relief of edema. There is no reason why the leg should not be elevated and no reason against its being moved, the proximal end of the thrombus being within the pelvis and little influenced by such factors. Indeed, why worry about casual exercise when the patient must practice daily the athletic feat of using the bed pan? So the foot of the bed should be raised six inches and the swollen leg elevated still more upon an inclined plane or in a sling. A couple of pillows may be placed under the shoulders and head but the body should not be bent by raising the upper half of the *adjustable bed*. Under these conditions, the leg will have freedom of motion and should not be covered with ice bags. Beyond the fact that heat usually brings more comfort—if the thigh is painful—the ice bag delays the flow of blood rather than hastens it and occasionally inflicts a frost-bite. The old custom was to sit the patient up in bed, apply ice to the flaccid legs and wait for swelling and fever to subside. Whether the existence of fever is actually an important consideration is unknown to the writer. When present, it may be expected to disappear with the edema. Under the system here described, as edema disappears, active exercise of the leg or legs in bed is begun and should be continued, with assistance if necessary, for some days before the patient is encouraged to get up. Then, with bandages applied up to the knees, he begins to walk, going to bed between his early attempts. From this time on, the use of the legs *must* slowly and regularly be increased. A return of edema *is of course* a sign of too much dependence and too little elevation. Standing, or sitting with the legs dependent, encourages swelling. Muscular exercise diminishes it.

Should pulmonary *infarction* occur, the question of dividing a great vein proximal to the thrombus comes up. For those who have not as yet been treated by elevating the lower half

bolism The presence of heparin not only delays coagulation but prevents adhesion of the thrombocyte, that is, it completely abolishes thrombosis Unfortunately, the treatment is very expensive, and is certainly not yet available for general use One may perhaps depend upon Bancroft's clotting index as a measure of the threat of thrombosis, taking special precautions or using an anticoagulant drug for the case in which thrombosis seems imminent For an account of the test, Bancroft's writings should be consulted His formula is shown below

$$\frac{\text{Prothrombin (1)} + \text{Fibrinogin (0 5 to 0 7)}}{\text{Antithrombin (1)}} = 0\,5\text{-}0\,7$$

An index of over 1 points to a tendency to thrombosis and the need of preparing the patient by the use of a high protein, low fat and carbohydrate diet Whether or not this diet is given, sodium thiosulphate in ten per cent solution is administered intravenously for three consecutive days in a daily dose of ten ccm

The Treatment of Established Thrombosis must always hurry the return of blood from the legs and pelvis A solid thrombus, it must be supposed, occupies the upper femoral and more or less of the iliac vein But is a loose, detachable, propagating clot present at the proximal end of the thrombus? In the usual outspoken phlegmasia alba dolens it is very rarely present in the occasional quiet, barely noticeable and perhaps incompletely obstructing thrombophlebitis it may well be However, the only proof of its existence is the occurrence of a pulmonary infarction due to a nonfatal embolus Therefore, unless one believes that the common iliac vein or vena cava should be divided rather often on suspicion (!) the presence of the propagating process must be ignored and all efforts directed against its *formation* It isn't the patient's turning over in bed which should be blamed for the fatal embolism, but the presence of the detachable embolus, and

the best practical defense against the formation of the embolus is elevation of the lower limbs.

Elevation of the legs has two objects: the hurrying of a collateral stream past the proximal end of the thrombus, to prevent the growth of the fragile clot which a slow stream encourages, and the relief of edema. There is no reason why the leg should not be elevated and no reason against its being moved, the proximal end of the thrombus being within the pelvis and little influenced by such factors. Indeed, why worry about casual exercise when the patient must practice daily the athletic feat of using the bed pan? So the foot of the bed should be raised six inches and the swollen leg elevated still more upon an inclined plane or in a sling. A couple of pillows may be placed under the shoulders and head but the body should not be bent by raising the upper half of the adjustable bed. Under these conditions, the leg will have freedom of motion and should not be covered with ice bags. Beyond the fact that heat usually brings more comfort—if the thigh is painful—the ice bag delays the flow of blood rather than hastens it and occasionally inflicts a frost-bite. The old custom was to sit the patient up in bed, apply ice to the flaccid legs and wait for swelling and fever to subside. Whether the existence of fever is actually an important consideration is unknown to the writer. When present, it may be expected to disappear with the edema. Under the system here described, as edema disappears, active exercise of the leg or legs in bed is begun and should be continued, with assistance if necessary, for some days before the patient is encouraged to get up. Then, with bandages applied up to the knees, he begins to walk, going to bed between his early attempts. From this time on, the use of the legs must slowly and regularly be increased. A return of edema is of course a sign of too much dependence and too little elevation. Standing, or sitting with the legs dependent, encourages swelling. Muscular exercise diminishes it.

Should pulmonary infarction occur, the question of dividing a great vein proximal to the thrombus comes up. For those who have not as yet been treated by elevating the lower half

of the body, elevation should first be tried But should infarction occur while the patient is being exposed to the favorable effect of elevation, the answer is difficult It has already been explained that embolism from an outspoken, completely obstructing femoro iliac thrombophlebitis is unusual It is far more common when swelling is slight or absent and a considerable current is able to flow past the thrombus Should such a condition be found upon exploration at the groin, opening the common femoral—the current from the various entering branches being controlled—may permit extraction of the insecurely fixed clot by gentle suction The femoral and its various branches should then be divided However, the indications for *this difficult* procedure or even a *direct attack* upon the iliac vein within the pelvis are not yet clear Approach to the left common iliac vein is best made transperitoneally, to the right, probably extraperitoneally from the direction of the iliac crest and right flank But now the operator must determine the upper limit of the thrombus, which may be very soft Unless he is fortunate enough to have divided the vein above it, he must still open the vessel and suck out the clot Possibly the approach to all iliac thrombi had better be made, as in arterial embolism, through the common femoral

Thrombophlebitis in the Prostatic and Uterine Veins—Thromboses found at autopsy in these vessels have indicated them to be a source of fatal pulmonary embolism, especially when no other source is discovered Probably thrombosis in the plexus of veins draining the prostate or the uterus is able to progress through the hypogastric vein into the common iliac, obstructing the return from the leg and causing phlegmasia alba dolens Indeed such must be an occasional cause of that disease But may a long, nonobstructing, loose clot spring from a prostatic or uterine vein and may it break off leaving only the parent thrombus deep in the pelvis? That seems not to be known There is certainly no way of identifying such a process during life One can only hope that the routine treatment intended to forestall a femoro iliac thrombophlebitis will prevent its occurrence

thrombosis is still active—is even perhaps extending—and that a propagating thrombus may well be growing up the femoral vein. There is all the clearer indication for femoral division in that experience has shown such treatment to cure the disease with remarkable rapidity. Of course it removes the danger of pulmonary embolism. One might suppose that division, even below the profunda femoris, would cause, for a moment at least, cyanosis of the foot. Such is not the case. The foot, which is usually a little cold, becomes warmer and even perhaps pinker than the other. In other words, a peripheral vasodilatation occurs.

Division of the femoral is performed under local infiltration with procaine or spinal anesthesia. A ten to twelve cm. (four to five inch) oblique incision parallel to the inguinal ligament and about three cm. (one inch plus) below it is satisfactory. The great saphenous vein is first found and is not disturbed. It actually is the best guide to the femoral. With retraction upward and downward upon the parts superficial to the aponeurosis, this layer is divided peripherally from the saphenous opening, exposing the common femoral just proximal to its division. Perhaps two cm. of the superficial femoral is isolated just distal to the profunda, silk ligatures are tied, the upper just below the profunda, and the vein is divided between them. This gives a good cuff both proximally and distally. If a segment is to be excised, a considerably longer exposure of the vein is needed. Should the propagating thrombus actually be encountered, it had better, as explained in the treatment of femoro-iliac thrombophlebitis, be extracted by suction and the vessel divided. The wound is closed in layers with fine silk and requires only a local dressing.

Following femoral division or resection, the patient remains in bed, the foot of which is kept elevated four to six inches, for a week or for such a period as the surgeon feels is required for the healing of the wound. The thrombosis is no longer important. Bandaging the lower leg is advisable when walking is begun but a normal life can soon be resumed. In the writer's experience, cyanosis is not afterwards noticeable, but a little

tempted The difference between the two legs is usually very clear to the patient Not only is this test useful in diagnosis but in following the progress of the disease, for with healing of the thrombosis and the re establishment of a normal circulation, the sign disappears

Treatment varies with the stage at which the disease is first seen If the patient has but just complained of lameness and now for the first time exhibits a slightly swollen, bluish foot and soreness on forced dorsiflexion of the foot, conservative treatment should be tried The lower end of the bed is raised, on blocks, four to six inches above the upper, and the affected leg is placed on a soft pillow A large cradle is used to keep the bedclothes off the feet No restriction is placed on moving the leg, but on the other hand no attempt is made at this time to exercise it A couple of pillows may be placed under the head and shoulders If the patient is able easily to manage the bed pan for defecation he should do so If not, he had better get up once a day (applying a semi elastic bandage from toes to knee) and use the bedroom cabinet

Elevation in bed is maintained for at least ten days or until all signs of soreness on forced dorsiflexion of the foot and all edema have disappeared The next four or five days are spent gently exercising all the muscles of the leg in bed For it is held that such exercise should precede getting up and that a propagating thrombus need not at this time be feared Then the patient begins to get up, wearing, when he first walks, a semi elastic bandage from toes to knee He feels his way along, walking a little and again elevating the leg, gradually increasing the periods of use and shortening those of rest If no swelling or blueness of the foot is noticed, an active life is resumed, the whole period of treatment having been three or four weeks

If, on getting about, swelling and cyanosis recur or if the patient is first seen when he (or she) has already, during several weeks, undergone successive periods of elevation, apparent recovery, and a return of the original signs, the femoral vein is divided distal to the profunda For it is reasoned that

sociated with a reddened, tender skin, local edema, and some induration. The redness is sometimes such as to suggest that a lymphangitis is present, and, moreover, the brownish discoloration which often afterwards remains does not always follow exactly the course of the vein. Perhaps the lymphangitis causes the thrombosis (Figure 25 shows how nearly the lymphatics are related to the superficial veins), in which case the common epidermophytosis of the feet may be a factor. In any case, such infection as is present rarely leads to suppuration. The process tends rather to become chronic, the thrombus remaining fixed in the vessel with little change over a period of several weeks. In the end, a combination of softening and organization occurs, by which the lumen of the vessel is restored, and it regains something very much resembling its previous state. But once having been thrombosed, a varicose vein is always liable to this accident, which is a sufficient reason for dealing radically with the process.

Palliative Treatment.—There are two ways of making the thrombosis of varix last a long time: the first is to go about without an elastic bandage, the second is to go to bed, sitting up with the legs outstretched in a horizontal position. Both methods keep the venous stream slow and encourage continued thrombosis. By contrast, the process is made to disappear by applying an elastic bandage and leading an active life, or, rather more effectively, by elevating the feet above the head and so remaining in bed. The first of these last two methods is especially useful when thrombosis is confined to the lower leg so that an elastoplast bandage can firmly be applied from the toes to the knee, that is, up to a point well above the level of the process. Ten days of such treatment—elastic pressure hurries the venous stream—often causes the thrombus to disappear but of course leaves the way open for a recurrence. Elevation of the leg in bed acts almost more quickly and has the advantage that it is equally successful when thrombophlebitis is present in the thigh. Indeed, when the process has threatened to reach the saphenous opening, it is the only satisfactory treatment.

edema on hard usage of the leg may be present for some months A brief note upon two typical cases of deep thrombosis in the lower leg will be found on page 216 A case in which femoral division was practised will be found below among a group illustrating diagnostic problems

Thrombophlebitis in Varicose Veins—The fibrosed, unhealthy state of the varicose vein's wall, associated with a feeble or reversed current, is explanation enough of the common thrombosis of varix One may presuppose infection in tissues of lowered resistance, or one may believe that the lining of a dilated, pocketed varicose vein actually cracks under heavy back pressure, since pressures of arterial height have been recorded on coughing or straining The wonder is, not that thrombosis occurs sometimes, but that it does not occur always It usually appears near the knee, more often below than above and in a prominent dilated vessel or pocket From its point of origin it progresses erratically upward, as a rule, and for an uncertain distance, but once half way up the thigh it is likely to reach the saphenous opening Beyond this it almost never goes, that is, it does not grow into the femoral either as a solid occluding thrombus or as a propagating clot threatening embolism

The failure of thrombosis in a varicose vein to invade the femoral is a good example of the ending and healing of thrombosis where it encounters a strong blood stream The writer knows of only two instances of pulmonary infarction caused by thrombophlebitis in varix Both of these were successfully treated by high division of the great saphenous Apparently the best reason for the solid attachment of a thrombus in the great saphenous vein is again the unhealthy state of the vein's wall This is in accord with the general principle already laid down, that the more outspoken the thrombophlebitis the less the danger of embolism The dependent position of the leg—most of the time—has little to do with the confinement of the process to the varicose vein, else the subject of thrombophlebitis had better never lie down

The thrombophlebitis of varix is occasionally acute and as

sociated with a reddened, tender skin, local edema, and some induration. The redness is sometimes such as to suggest that a lymphangitis is present, and, moreover, the brownish discoloration which often afterwards remains does not always follow exactly the course of the vein. Perhaps the lymphangitis causes the thrombosis (Figure 25 shows how nearly the lymphatics are related to the superficial veins), in which case the common epidermophytosis of the feet may be a factor. In any case, such infection as is present rarely leads to suppuration. The process tends rather to become chronic, the thrombus remaining fixed in the vessel with little change over a period of several weeks. In the end, a combination of softening and organization occurs, by which the lumen of the vessel is restored, and it regains something very much resembling its previous state. But once having been thrombosed, a varicose vein is always liable to this accident, which is a sufficient reason for dealing radically with the process.

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Local applications are not essential If any are used, heat is preferable to cold It encourages hyperemia, brings comfort and presumably discourages further thrombosis, whereas the traditional ice bag delays the circulation, devitalizes the tissues, and encourages an extension of the thrombus The only good which can be said of the ice bag is that it is often comforting, quite as much so as the hot-water bottle

Curative Treatment—To check thrombosis, shorten the patient's disability, and prevent recurrence of the disease nothing can compare with resection of the great saphenous vein at its entrance into the femoral The only contraindication to such treatment is the presence of a thrombus in the upper part of the vein However, this contraindication is not absolute and in many cases it is difficult to decide whether or not thrombosis is actually present at the saphenous opening Should it be determined beforehand, because of the presence of a thickened vein and local tenderness, that such is actually the case, the patient is subjected to the routine elevation of the foot of the bed Within ten to fourteen days, the local thickening and tenderness will usually have disappeared, indicating that the thrombus has been organized or liquefied. As one follows a superficial thrombosis one is aware that the vein becomes continually softer, so that it is less and less easily palpated From being a solid cord, the size of a lead pencil, it will often in a space of ten days, more or less, take on the character of a soft, barely palpable vessel Its wall always becomes thickened, but not to such a degree as to suggest that the whole thrombus is organized Apparently the most clot like portion is liquefied and carried away

Once the upper saphena magna is again open, it may well be resected Opinion will naturally be divided as to when the operation should be performed, but the likelihood of recurrence is such that a high division had better be made sometime Why not, therefore, resect the vein at once and avoid a subsequent hospitalization?

If exploration at the groin unexpectedly finds a thrombosed saphenous vein, it has been the writer's practice to pass two

ligatures under it at the point where it is exposed and divide the vessel between the ligatures. In so doing the operator should disturb it as little as possible but actually the danger of detaching a part of the thrombus is very slight. For not only is division made an inch from the femoral but a propagating clot very rarely grows into the latter vein.

Should the thrombus end proximally below the saphenous opening, the operation of dividing the great saphenous vein is performed in exactly the same way as for uncomplicated varix. One should have in mind, however, that almost every superficial thrombophlebitis is associated with some involvement of the perivenous lymphatics and some degree of lymphadenitis at the saphenous opening. Any suggestion of enlarged inflamed nodes is therefore a signal for especial care lest the glands be disturbed and spill infection into the wound. However, such an infection never seems actually to suppurate. Following high resection of the great saphenous, the thrombosis clears up very rapidly. The varicose veins as well are usually cured.

Thrombophlebitis in Nonvaricose Superficial Veins.—The form known as *phlebitis migrans* has been described in Chapter III as a complication of thrombo-angiitis obliterans. There it takes on its most typical appearance. However, a disease, seemingly in other respects identical, does occur in those who are not sufferers from Buerger's disease. In some of these there is a tendency to recurrence throughout life. Without any obvious cause, a stretch of vein, an inch or two in length, almost invariably upon the surface of the lower leg, becomes solid, thickened and slightly tender. In this state it remains for a week or two, and then, as it softens and apparently is restored to something very much like its normal condition, another area of thrombophlebitis, considerably proximal to the first, appears. The first may occupy the region of the ankle, the second, the upper calf, and a third, perhaps, the lower thigh. Apparently there is little tendency to embolism and continued use within an elastoplast bandage is about as successful as any other form of treatment. Whether the dis-

case would promptly disappear (on any one occasion) if the leg were subjected to continuous elevation is unknown to the writer It is usually treated by only partial rest and elevation and has exhausted everyone's patience before it ceases to break out A fairly typical example, in which high division was finally used, will be found included with several other case reports below

The superficial thrombophlebitis which does not take the form of phlebitis migrans is actually more freakish and unaccountable than any other It occurs most often perhaps in locally dilated veins which, however, are not a part of a varicose saphenous system Local chafing, as in horseback riding, has been known to bring it on Exposure to unusual cold, a severe bout of coughing, a trivial injury of any sort may occasion thrombosis It seems to have the faculty of hanging on for considerable periods, and if not promptly cured may at any time quietly march out of a small radical into the main stem of the great saphenous and from somewhere in the course of that vessel give off an embolus Altogether it is difficult to know whether or not to fear it For in most cases such a thrombosis must heal with little to show for its presence It must also be admitted that among adipose women, in whose fat a local thrombosis is not easy to identify, the diagnosis between it and a local lymphangitis or mild cellulitis must often remain doubtful

Treatment —Following the general rule of treating thrombosis, the superficial form, if only suspected of being present, should be treated by elevation—of the legs above the head—over a period of perhaps ten days This period is set to offer a safe margin for thrombophlebitis not easily palpated As a matter of fact, the writer has seen it disappear completely from a vein near the ankle in less than a week But if, as so often happens in this and other sorts, the thrombosis has already persisted for several weeks when first seen, it should receive more radical treatment That is to say, the parent vein, usually the great saphenous, should be divided at the saphenous opening, after which the process is soon healed

Cases Illustrative of Various Sorts of Thrombophlebitis—and Their Treatment

The reader may, if he likes, come to a conclusion as to the nature of the disease in these cases and plan the treatment. He will perhaps prefer his own plan to that actually used.

Case 1.—R.C., an athletic man, forty-eight years of age, while playing tennis six weeks before coming under observation, gave his left leg an unusual wrench. He suffered considerable pain but was able to continue play. He must have torn a muscle (?) in the upper calf for he noticed considerable local ecchymosis in the course of the days following. Nevertheless he kept about for three days, his lower leg somewhat swollen and painful. Then he took to his bed, with such relief that at the end of a week he thought himself fit to get about again. However, on getting up he soon noticed that the left ankle was swollen, the foot bluish. Again he went to bed. for nine days this time, and again on getting about the foot became cyanotic, the lower leg swollen.

When he came under observation, he had gone to bed for the third time and had had six days of it. The left leg looked normal but the foot felt cool to the touch as compared with the other and turned a little blue when hung for a couple of minutes out of bed. There was a slight feeling of soreness and tension, behind the knee, on forced dorsiflexion of the left foot. On the following morning, the patient applied a bandage and went to his office, but when seen that afternoon, the foot was blue and the marks of the bandage showed clearly upon the swollen leg.

Diagnosis.—Rupture of muscle (and vein?). Thrombophlebitis of deep veins of left lower leg. Immediate division of femoral vein advised.

Operation.—The femoral vein, which appeared normal and contained no thrombus, was divided just below the profunda. A segment excised was not remarkable. Immediately (on the operating table) the left foot, hitherto colder than the other, became equally warm and if anything a little pinker in color.

The patient left the hospital in a week wearing a semi elastic bandage In three weeks he had begun to exercise as usual At first the ankle would be found a little swollen each night but would be normal in the morning After six weeks he gave no further attention to his leg

Case 2—B S N, a woman, fifty two years of age, had suffered during her sixth pregnancy, nine years earlier, from an inflamed vein upon the inner side of her left thigh This troubled her during the last few months of her pregnancy During delivery (placenta previa) she lost much blood, and two weeks later developed a milk leg This came on with terrific pain and lasted for two months The leg had never been the same since, swelling at the ankle when she was for long hours on her feet

Four months after delivery and the onset of the milk leg, the patient suffered what seemed to be a pulmonary infarct (severe pain beneath the left breast but without hemoptysis) This was repeated, without obvious reason and in a far more severe form, eight years later—severe thoracic pain, fall of blood pressure and prostration Soon after recovery from this episode, the writer saw the patient and found a slightly darkened area upon the inner face of the upper left calf, the remains, he thought, of a recently healed great saphenous thrombosis No actual varicose veins As the patient had been up and about very little, he suggested seeing her again when she had been more on her feet but he only saw her six months later after her third and almost fatal pulmonary embolism

This embolism, which was marked by agonizing pain behind the sternum and a violent fall of blood pressure, was preceded only by a little patch of redness and soreness upon the inner side of the calf (in the region previously thought to be the scene of a saphenous thrombosis) This was remembered afterward In the meantime, the E K G was found to be consistent with a pulmonary embolism and the X ray of the thorax with a pulmonary infarct

Upon recovery, the state of the leg was the following Redness and soreness of the calf had disappeared. However,

higher up, on the thigh, a faint discolored patch over the course of the great saphenous vein was noticed. But at the same time there was typical discomfort back of the knee on forced dorsiflexion of the foot.

Thus the question was raised whether the last embolus had come from the saphenous system (evidence of recent thrombosis) or from the deep veins of the calf (dorsiflexion sign). More than this, there was, in the background, the old femoro-iliac thrombosis, so that the embolus could conceivably have come from the iliac region. It was decided to divide the great saphenous and to examine the femoral, with the idea of dividing it unless it contained a thrombus. It seemed desirable to operate promptly when the state of the vessels was likely to give a clue to the recent course of events.

At operation, the great saphenous was found whitish, thick-walled, evidently the scene of an old thrombophlebitis (which might have been secondary to the original milk leg). There were enlarged, juicy lymph nodes about the saphenous opening as evidence of a recent process. The femoral artery and vein were embedded in fibrous tissue and were dissected apart with some difficulty (common result of old phlegmasia alba dolens). The vein contained no thrombus. It was divided distal to the profunda, between silk ligatures.

Immediate result, normal color and warmth of left foot. Normal healing. Unaccountable weakness of extensor muscles of both thigh, leg, and foot, from which recovery was gradual but complete. Ankle and foot afterwards swelled less than formerly. Last report, only six months after operation, showed continued good health. No one will ever know, of course, from which vessel the emboli had come, the great saphenous or a deep vein in the calf, but the episode is unlikely to be repeated.

Case 3.—E.D., a woman, sixty-two years of age, the mother of two children. Typhoid fever, rheumatic fever, and malaria in youth. Following her first pregnancy, she had noticed enlarged veins on the inner face of the thigh and outer side of the left leg below the knee. No typical varicosity of the great saphenous system.

About a month before being seen by the writer, the patient noticed an area of redness and soreness over the enlarged veins outside and below the left knee, together with some crampy feeling in this region which came on suddenly at night and promptly disappeared She kept about under considerable discomfort for four days, at the end of which time she consulted her physician who made a diagnosis of "phlebitis" and put her to bed There she remained, sitting up most of the time, her leg on a pillow, for two weeks The area of redness in the meantime subsided

At the end of her two weeks in bed, the patient experienced a sudden epigastric pain and a desire to defecate (a common premonitory sign of pulmonary embolism) At stool, she was stricken with a sharp, severe pain in the right chest, becoming weak, breathless and apprehensive With the aid of an oxygen tent and cardiac stimulants, she survived As soon as possible, the left leg was elevated and the head lowered Both E K G and X ray confirmed the diagnosis of pulmonary embolism and infarction

On recovery from the embolism, the patient was found to present no asymmetry of her legs, no blueness or swelling of the left foot The veins of the left leg, lateral to the knee, were a little prominent as were those of the inner face of the thigh No tenderness or masses, but at the groin, in the region of the saphenous opening, was a pencil shaped, slightly tender mass, three cm in length which, from its situation, might have been either a thrombosed saphenous vein or a group of slightly inflamed lymph nodes Under several days' observation this mass disappeared

There was no discomfort on forced dorsiflexion of the foot

Diagnosis —Superficial thrombosis, probably in the great saphenous vein (history, presence of enlarged veins and tender mass at saphenous opening) but possibly in the lesser saphenous (position of original soreness and redness)

Operation —Division of great and lesser saphenous veins The lesser saphenous was found sclerosed and irregularly distended Near the popliteal, it was small and not thrombosed

It was not believed to have been the source of embolism but was divided.

The great saphenous vein at the groin was straight and thick-walled. About a large branch, entering it laterally, the tissues were adherent. A vague mass of lymph nodes was present and was not disturbed. Evidence of recent inflammation in the upper saphenous and a large lateral branch being clear, and no thrombus being now palpable, the great saphenous was resected, from the femoral downward for over an inch (three cm.). Pathological examination revealed an organizing thrombus in the resected portion! A propagating thrombus must have grown from the great saphenous into the femoral while the patient reclined in bed for two weeks.

The patient made an excellent recovery. The left leg swelled a little at first at the ankle when the patient spent much time on her feet—actually less than before the operation.

Case 4—R.E.F., a man, forty-five years of age, was first seen complaining of a sore spot upon the inner face of his calf, a little below the knee. Six years earlier he had noticed, at a time when he had been going up and down stairs a good deal, a sudden pain in the back of his left calf, which then became sore to the touch. He never really gave in to the illness which [illegible] he [illegible] end of which time bl[illegible] began to spit up [illegible] He was at first thought to have tuberculosis, a diagnosis afterwards contradicted by an expert on that disease. No one, however, had connected the state of the leg with that of the lung. He had since remained well until his present illness.

When seen by the writer three days after the appearance of the new area of soreness, there was noticed a little reddish streak overlying an obviously thrombosed vein. In those three days, the thrombosis had risen slightly until it reached the natural crease just below the knee, rather posteriorly. The leg was shaved and an elastoplast bandage applied from toes to knee. The patient was directed to go about as usual. Eight days later the region of the thrombosis was no longer sensitive and seemed to have healed but on the following day, the

About a month before being seen by the writer, the patient noticed an area of redness and soreness over the enlarged veins outside and below the left knee, together with some crampy feeling in this region which came on suddenly at night and promptly disappeared. She kept about under considerable discomfort for four days, at the end of which time she consulted her physician who made a diagnosis of "phlebitis" and put her to bed. There she remained, sitting up most of the time, her leg on a pillow, for two weeks. The area of redness in the meantime subsided.

At the end of her two weeks in bed, the patient experienced a sudden epigastric pain and a desire to defecate (a common premonitory sign of pulmonary embolism). At stool, she was stricken with a sharp, severe pain in the right chest, becoming weak, breathless and apprehensive. With the aid of an oxygen tent and cardiac stimulants, she survived. As soon as possible, the left leg was elevated and the head lowered. Both E.K.G. and X-ray confirmed the diagnosis of pulmonary embolism and infarction.

On recovery from the embolism, the patient was found to present no asymmetry of her legs, no blueness or swelling of the left foot. The veins of the left leg, lateral to the knee, were a little prominent as were those of the inner face of the thigh. No tenderness or masses, but at the groin, in the region of the saphenous opening, was a pencil shaped, slightly tender mass, three cm. in length which, from its situation, might have been either a thrombosed saphenous vein or a group of slightly inflamed lymph nodes. Under several days' observation this mass disappeared.

There was no discomfort on forced dorsiflexion of the foot.

Diagnosis.—Superficial thrombosis, probably in the great saphenous vein (history, presence of enlarged veins and tender mass at saphenous opening) but possibly in the lesser saphenous (position of original soreness and redness).

Operation.—Division of great and lesser saphenous veins. The lesser saphenous was found sclerosed and irregularly distended. Near the popliteal, it was small and not thrombosed.

usually arises from a propagating thrombus occupying the whole length of the femoral, is rarely if ever survived. The embolus is too long and too heavy. However, it seemed wisest to cut off both sources. Resection of the diseased vein appears also to be actually beneficial to the peripheral circulation (release of vasospasm?).

Case 3 represents almost certainly a pure great saphenous embolism and brings up the question whether the propagating thrombus had floated, before detachment, in the common femoral or belonged entirely to the great saphenous system, having been detached from some point in the lower thigh or calf. The former seems the strongest probability in spite of the extreme rarity of embolism from the femoral end of the thrombosed great saphenous vein. That is, a propagating thrombus almost never forms and hangs in the strong femoral current when the saphenous vein is thrombosed. Here, however, the patient had remained in bed for two weeks, suffering from what must have been at first a very local superficial thrombosis, much of the time reclining and thus creating conditions favorable to propagation of a thrombus.

In Case 4, which should have been subjected to operation two months earlier, adhesive bandaging was given more than a fair trial. Apparently the state of the patient's vein was the important factor. He showed, according to Bancroft's index, no thrombophilic tendency, and his disease was quickly abolished by high division of the great saphenous. It is fairly typical of the phlebitis migrans type.

As a group, these cases illustrate the utter harmlessness, from the point of view of obstructing the venous return, of dividing a great vein when disease is present in its peripheral portion. The *benefit* to the circulation is perhaps due to the interruption of undesirable impulses passing along the vein, in which direction is not clear. The matter is perhaps analogous to arterial resection in the presence of a local plug. The cure of the thrombosis itself is always striking. After division or resection of the vein, the peripheral thrombophlebitis can almost be ignored.

bandage still being in place, a fresh area of thrombosis about two inches in length appeared just above its upper limit Fresh adhesive bandages were therefore applied up to a higher level, but in a few more days, the thrombosing process jumped again, this time to the mid thigh There it remained stationary, the lower areas being healed and free from soreness The patient continued to use the adhesive bandage and during the *following month, when in New York, consulted Dr Bancroft* who found his clotting index normal and advised high division of the great saphenous vein Some three months after the onset of thrombosis, the process still being active, the patient consented to operation

The Diagnosis was thrombophlebitis of the (nonvaricose) saphenous vein—phlebitis migrans type

Operation—The great saphenous vein was resected in the usual way at its junction with the femoral It was not diseased at this point Several days later his rather badly diseased tonsils were removed

The immediate result of operation was a rapid disappearance of the thrombophlebitis without further treatment The patient has remained well

Comment

These four cases are presented, not to show that operation is the only treatment for thrombosis, but to illustrate how it may be used when a fatal embolism is feared. Case 1, a typical *example of a persistent thrombophlebitis in the deep* veins of the calf muscles, would have been treated, if seen early, by elevation alone Yet after six weeks of shillyshallying, an operation offered not only the quickest, surest means of cure but an almost certain safeguard against embolism

Case 2 was a complicated one, for it presented suggestive evidence of thrombosis in both the superficial system and the deep veins of the lower leg Probably the embolisms which had already occurred came, however, from the great saphenous stem This conclusion is reached principally because they were not fatal Embolism from the deep peripheral veins, which

slightly enlarged arm is faintly cyanotic. It is impossible to say as yet whether such a condition is likely to persist. If so, an axillary periarterial sympathectomy or a sympathetic ramisection would presumably be curative.

Pulmonary Embolism

In the foregoing account of thrombophlebitis, reference has repeatedly been made to the varieties most liable to the formation of an insecure propagating thrombus and the subsequent detachment of an embolus. A brief summary of these matters and a reference to promising methods of treating minor pulmonary embolism are included in the following paragraphs.

That there should be formed in a vein a floating, waving, friable clot of sufficient thickness and length to plug the pulmonary artery, it is necessary that a stretch of some ten to twelve inches in a medium- to large-sized vein should be available proximal to the thrombus. Such, of course, is found in the superficial femoral, the external, and the common iliac. A propagating clot may project from a vein of the deep calf or popliteal space into the femoral, from the femoral into the external and common iliac, from pelvic veins through the hypogastric into the common iliac and vena cava, and from the common iliac into the vena cava. Doubtless other situations might be suggested, but the femoral system and the veins of the pelvis, in both of which the venous stream can so readily be retarded, seem to be the most available sources of embolism. Particular spots anatomically favorable to thrombosis are present in certain parts of these vessels, namely in the upper part of the deep calf, the region of the groin, and the deep pelvis.

The large, fatal embolus is often a foot long and may show at one end the facet left by its detachment from the parent thrombus. Sometimes a number of separate emboli are found in the pulmonary artery or one of its main branches as if smaller fragments had been detached one after the other. Such great masses are usually rapidly fatal, but rarely the embolus

Thrombosis (by Effort) of the Axillary Vein

This sort of thrombosis is rare and is evidently a very special form It is seen almost always in active individuals, of the third, fourth, and fifth decades, who have recently made with one arm or the other—the right as a rule—some unaccustomed effort Hence the name "thrombite par effort" bestowed upon it by the French Those who have most carefully studied the accident believe that when the arm is abducted, and during expiratory engorgement of the axillary vein, the costo coracoid ligament indents the vessel (Lowenstein) or the subclavius muscle actually stretches the vein's wall at a certain point and injures a particular large valve (Gould and Patey) In any case, it is the arm most actively strained which exhibits the thrombosis From the upper axillary vein, the process extends downward for a variable distance toward the elbow

The usual history tells of some rather strained exertion with the arm elevated Almost at once pain or swelling of the arm sets in, and during the next day or two the whole limb becomes edematous and somewhat blue The superficial veins are apt to stand out, particularly over the shoulder The axillary vein itself can be felt as a cord or elongated lump

Treatment consists of rest in bed and elevation of the arm upon pillows Fixation is unnecessary Under these conditions the discomfort and swelling disappear very much more rapidly than is the case with a femoro iliac thrombosis In ten days to two weeks, the arm will usually have regained its normal appearance, save perhaps for some enlargement of the veins about the shoulder There may also be left some temporary stiffness—no permanent after effects Embolism does not seem to be a danger

As a rare complication of an effort thrombosis, the individual may be left, as in other forms of thrombosis, with some degree of vasospasm, even a mild causalgia like state The writer has seen one such case The individual suffers a pain, something like that of intermittent limp, on exertion, and the

haps a mild lymphadenitis at the saphenous opening. A deep thrombosis in some of the great plexuses among the muscles of the calf gives a characteristic story and often is betrayed by subjective discomfort behind the upper calf on forced passive dorsiflexion of the foot. The detection of a femoro-iliac thrombophlebitis is less easy. The outspoken sort—phlegmasia alba dolens—causes the familiar painful or uncomfortable swelling of the whole lower limb, but this sort seldom causes embolism, and if it does, the embolus comes from the external or even the common iliac, a situation, which, except under the most unusual circumstances, must be regarded as out of reach. The quiet, incompletely obstructing, embolism-threatening sort is practically undetectable—there may be no swelling or cyanosis and even no discomfort or tenderness over the femoral vessels at the groin. As for thrombosis in the depths of the pelvis, which is presumed to exist, it is absolutely silent.

An account of how these various processes can best be treated and how, in the presence of some of them, the patient can be protected against further embolism, has already been given. In many cases, effective preventive treatment can be established.

The treatment of the fatal type of pulmonary embolism by Trendelenburg's operation, performed upon the unconscious and seemingly moribund patient, that is, exposing the heart, opening the pulmonary artery and sucking out the embolus, should be studied in the writings of Trendelenburg and, more recently, of Nystrom. By a perfect cooperative technique, possible only in first-rate hospitals, an occasional life can be saved, as Nystrom relates, yet a more promising field is probably offered by treatment designed to prevent thrombophlebitis, or, if thrombosis has already occurred, to cut down the incidence of embolism.

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will float for some days in the artery without being forced into one of its main stems or causing more than a partial obstruction of the pulmonary circulation As a rule, however, the patient is struck down by breathlessness and an agonizing oppressive discomfort or pain in the mid line behind the sternum Breathing is violent, the accessory muscles of respiration are called upon, and cyanosis is succeeded by pallor, feeble breathing and death The first sign of trouble may be a vague abdominal discomfort and a desire to defecate Occasionally the substernal pain and respiratory distress almost exactly imitate coronary infarction, so that only by the aid of an electrocardiogram can a distinction be made between the two

Small emboli, causing pulmonary infarction, seem to be of two sorts first, fragments of embolus detached from a large thrombus and, second, entire loose propagating clots from vessels of moderate size The first sort are likely to be followed by the fatal detachment of a large embolus The second cause only the familiar infarction, leaving an area of dullness and diminished respiration in one lung or the other and a patch of decreased aeration detectable by the X ray As a rule, infarction causes the patient to expectorate some blood and a pleuritic pain is usually present The seriousness of the situation depends upon the size of the embolus Minor infarctions are recovered from very rapidly but the larger ones are shocking and require cardiac stimulation and the use of the oxygen tent Naturally, the occurrence of infarction leads to a search for its source, in order to determine whether a vein can be divided proximal to the process, protecting the patient from further and perhaps fatal embolism

The search for the source of any embolus will lead to a study of the superficial veins of the leg, the deep veins of the calf, the femorals and iliacs The nature and course of thrombophlebitis in these various vessels has already been explained Evidence of a thrombophlebitis in the great saphenous system will usually be found in the form of a story of local tenderness and redness, the actually palpable thrombosed vein and per

ps a mild lymphadenitis at the saphenous opening. A deep rombosis in some of the great plexuses among the muscles f the calf gives a characteristic story and often is betrayed y subjective discomfort behind the upper calf on forced pasive dorsiflexion of the foot. The detection of a femoro-iliac thrombophlebitis is less easy. The outspoken sort—phlegmasia alba dolens—causes the familiar painful or uncomfortable swelling of the whole lower limb, but this sort seldom causes

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CHAPTER VII

ARTERIAL ANEURYSM ABNORMAL ARTERIOVENOUS COMMUNICATIONS

ARTERIOVENOUS ANEURYSM AND FISTULA

ARTERIAL ANEURYSM

AN ANEURYSM is a local dilatation of an artery It is the media which fails This is the muscular layer of all but the largest arteries Of these it is the heavy, elastic coat which confers such an astonishing combination of strength and elasticity upon the aorta and its main divisions The media of the great arteries gives way chiefly because of syphilitic infections Smaller vessels such as the femoral or popliteal suffer from aneurysm which is less apt to be syphilitic Arteries often and actively plied, as at the knee and groin, have a greater tendency than others to crack or bulge, and syphilis as a background is not required Of the great aneurysms, those of the aorta are the most common, those of the innominate and subclavian being next in rank Of the peripheral aneurysms, the popliteal is the best example and as a matter of fact, by far the commonest

The form of an aneurysm depends first upon whether a considerable stretch of arterial wall gives way or whether one particular spot weakens A diffuse weakening makes a fusiform aneurysm a local one, a saccular aneurysm However, time modifies these forms A saccular aneurysm, slowly enlarged, may so stretch the vessel, upon one side of which it first lay, that the original form of the artery is altogether lost Then the artery, becoming flattened, may see its lateral opening lengthened and expanded until finally the sac assumes an almost fusiform shape By contrast, a fusiform aneurysm may rupture, acquiring an almost saccular shape Such distinctions

are important chiefly from the point of view of treatment. That is, a true fusiform aneurysm can rarely be subjected to a plastic operation intended to restore a lumen, but a saccular aneurysm can sometimes be treated by one of the ingenious procedures of Matas, "restorative" or "reconstructive" aneurysmorrhaphy. Some arterial dilatations are so extensive that the whole vessel widens over a very long distance. In the aorta this is common enough, but the same thing may happen to practically all the great arteries of the body. The vessels of the legs, for instance, rarely become arteriosclerotic winding channels one to two centimeters in diameter. Such states as that are not considered here. It is proposed only to describe the subclavian and popliteal types of aneurysm, as representatives of those which affect the circulation of the limbs, to give some account of the tests intended to reveal the nature of the collateral circulation and discuss very briefly the standard methods of treatment.

Subclavian Aneurysm

This aneurysm, a representative of the great vessel type, is usually fusiform and presents itself as an expansile swelling which gradually appears above the clavicle at the root of the neck. If particularly large, it has been known to fill the space between the clavicle, scapula, and sternomastoid muscle, even lifting the clavicle and bulging into the axilla. It is more often right- than left-sided and nearly confined to males. Because of its proximity to the brachial plexus it is likely to be a cause of pain and weakness in the corresponding arm, and pressure upon the companion veins will result in venous congestion and edema. The radial pulse may be weak and delayed, the blood pressure in the corresponding arm lower than that of the opposite side. The natural course of the lesion is toward final rupture upon the surface at a point where the skin will already have become reddened and adherent. Before any surgical attack is made upon such an aneurysm, the state of the collateral circulation must first be studied. Actually there is no authoritative test for the efficiency of this circulation save by tem-

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Arterial Aneurysm

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the afferent artery can be controlled. The same thing should, if possible, be done for the efferent vessel, to check any retrograde flow into the aneurysm from this source. It should then be possible to open the sac widely. Bleeding from any entering branches must be stopped by pressure with the finger, and the branches closed by silk or chromicized catgut stitches taken across their mouths. Next, with circular stitches which take a good grip upon the inner coats of the aneurysm, the afferent and efferent orifices are closed from within. By a series of similar circular stitches, placed at short intervals from one end of the sac to the other, the whole of a small aneurysm can then perhaps be obliterated, but if the sac is larger and friable, so that the stitches will not hold, the wall of the aneurysm had better be infolded and made into a compact mass by mattress stitches. Much will depend upon how strong a retrograde flow is found to enter the sac from any branches encountered. The less such a flow, the easier and less bloody the operation. This of course is the same general plan which Matas uses in the treatment of any aneurysm of a smaller vessel which can not be reconstructed or restored. In the end, the controlling rubber tubes are released and if the obliteration has been thorough, all bleeding will be found to have ceased.

Popliteal Aneurysm

This is decidedly the most common peripheral aneurysm, comprising perhaps fifty per cent of all such lesions, the femoral and femoro-iliac (combined) coming next in frequency. It is usually of the fusiform type but may be saccular, representing what is almost a rupture of the artery at some one point. When still so small as to call no attention to itself by local swelling, the aneurysm sometimes causes coldness or numbness of the foot, perhaps an intermittent limp, that is, commonplace symptoms of an arterial deficiency. When it has grown larger, the actual tumor beneath the deep fascia of the popliteal space, which is so likely to interfere with the movements of the knee joint, often out of a clear sky calls atten-

porary occlusion of the afferent artery, since digital proximal compression, in the case of any but a small aneurysm of the third portion of the subclavian, is impossible The clavicle must be removed, the proximal portion of the aneurysm exposed, and a Matas or Halsted aluminum band closed about the entering artery If this is tightened just sufficiently to stop all pulsation in the aneurysm, the state of the circulation in the arm will become clear in the following day or two In the event that the arm and hand remain reasonably warm and pink, nothing more, for the moment, need be done, but the state of the sac will of course be watched If it becomes smaller, harder, and remains without pulsation, the aneurysm may be considered cured If the response is unfavorable, that is, if the hand and arm become white and cold, threatening gangrene, the closed wound should be reopened and the band, which has injured neither intima nor media, should be loosened just sufficiently to restore the circulation Even then, the development of the aneurysm may be checked or it may actually, by a process of thrombosis and organization in the periphery of the sac, be reduced in size Naturally, its subsequent course should be followed with care Even though the pulsation and enlargement recur, a collateral circulation will doubtless have developed

Operative Treatment—Should the application of a band have failed to control the aneurysmal pulsation or should pulsation and swelling have reappeared after a temporary improvement, more radical treatment will have to be tried This will usually mean some form of aneurysmorrhaphy, that is, infolding and obliteration of the sac, a method best adapted to preserving the collateral circulation

For aneurysms of the great vessels, the obliterative aneurysmorrhaphy of Matas is most likely to succeed and produce a permanent cure The region of the aneurysm is widely opened, the sac, with its afferent and efferent arteries, is as fully as possible exposed and the entering vessel especially is so well isolated that a piece of rubber tubing can be passed under it Thus by lifting on the rubber tubing the current in

others, and those interested in the subject will do well to study his writings.

The more simple yet reasonably reliable tests are the following:

1. Delbet's test. If the peripheral pulses distal to the aneurysm are absent, yet the limb is of good color and nutrition, the collateral circulation is almost certain to be dependable. The reason for this is clear enough. For if the main channels are obstructed or receive no arterial flow, the well-nourished limb *must* be getting its blood by collateral channels.

2. If the principal artery is compressed just proximal to the aneurysm and the peripheral parts fail to turn yellowish white but rather remain a reasonably pinkish white, the collateral circulation is sufficient.

3. If the principal artery is compressed as above and an oscillometer or even a blood pressure cuff applied to the peripheral part reveals some trace of pulsation, the collateral circulation is probably sufficient.

The more complicated and authoritative tests are the following:

1. Matas's flushing test (attributed by him to Moszkowicz) which is especially useful in the case of a popliteal aneurysm but which can also be used for any femoral lesion which is not too high. The artery immediately proximal to the aneurysm being compressed by the fingers or a mechanical device until the pulsation in the sac is abolished, and the leg being elevated, an Esmarch bandage is firmly applied from the toes up to the lower pole of the aneurysm. In this way the leg is emptied of blood. It is kept in this state for ten minutes in youngish persons, but for not more than five in elderly. Now, while pressure is maintained on the afferent artery, the Esmarch bandage is rapidly removed and any flushing (Reactive Hyperemia—Chapter I) must depend upon the efficiency of the collateral circulation. It is very easy to follow the descent of the resulting blush. If the collateral circulation is very competent, the flush will reach the toes in a few seconds to a minute or so. Usually it will go rather rapidly to the upper

tion to the expansile pulsating swelling If the sac cracks open at some point, the aneurysm may enlarge rather rapidly by a process of giving way, thrombosis, organization, and further stretching or rupture If the peroneal nerve is injured by pressure there will be some degree of toe- or foot drop Swelling and cyanosis of the foot and ankle are to be expected, but the most serious complications come from thrombosis within the sac If this process extends into the efferent arteries, the anterior tibial and the terminal branches, that is, the peroneal and posterior tibial, the foot will occasionally become gangrenous, especially in arteriosclerotics whose collateral circulation has not become abundant

Most popliteal aneurysms occur in middle life and for no apparently sufficient reason, though forced flexion followed by a violent muscular effort has sometimes been observed and syphilis is present in a small proportion of cases They are almost unknown in women, even though elderly females often suffer from arteriosclerotic disease of the arteries in their lower limbs The truth is that active use of the legs soon after the arteries have begun to stiffen rather than advanced arteriosclerosis seems to be the cause of most popliteal aneurysms Once the expansile pulsation of a good sized popliteal aneurysm is present, the diagnosis is clear At an early stage, however, when only signs of arterial deficiency are evident, diagnosis is very difficult The aneurysmal sac may be small or sufficiently thrombosed to prevent a deep pulsation from being appreciated, that is, if palpable at all, it will feel solid Yet all the while it may be responsible for a serious intermittent limp or impending gangrene Theis calls attention to the combination of very low peripheral skin temperatures with an excessive elevation of the oscillometer readings at the knee In case of doubt arteriography can of course be used

Tests of the Collateral Circulation—It is in the case of the aneurysms of the lower limbs that special tests of the collateral circulation are particularly needed The upper limb, in this respect, is far better safeguarded Matas has made a very thorough study of these methods, condemning some, accepting

tubing, the sac is opened from end to end and cleared of thrombi. All entering vessels are closed by stitches taken through the inner coats of the aneurysm. The sac is then closed from within by a series of circular stitches. If it is too large or too friable for such a step, it is infolded and matted

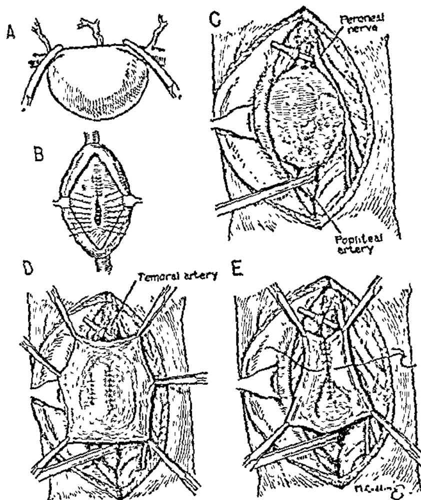


FIGURE 23. ANEURYSMORRHAPHY—after Matas. *A.* After Matas's sketch showing how most of collateral circulation can be controlled. *B.* Matas's plan of obliterative aneurysmorrhaphy. *C.* A popliteal aneurysm, showing the sac exposed (the perineal nerve has been compressed) *D.* The sac partly closed by aneurysmorrhaphy. *E* Further closure of sac with silk. The free edges may now be turned in and united by mattress sutures.

level of the ankle, after which it spreads to the toes in a hesitating, erratic, and patchy manner However, if the toes are colored pink in three minutes, the result may be considered satisfactory

Should the flushing time be considered too slow, the patient's leg should be subjected to measures intended to improve the collateral circulation These will include compressing the afferent artery for perhaps ten minutes at a stretch several times a day, and the usual vasodilating stimuli, especially the application of heat to the body Vasodilating exercises might cause rupture or thrombosis in the aneurysm Too much heat applied to the limb might be harmful though the leg should always be kept warm Massage, avoiding the sac itself, will be useful

2 The Application of a Matas or Halsted aluminum band—The rationale of this test has already been explained. The band should not injure the afferent artery and should be placed so close to the aneurysm as not to blot out any collateral arteries however small The various possible results of its application have been described under Subclavian Aneurysm If the peripheral part remains well nourished, the band may be left on with a reasonable hope of cure If the peripheral part does not seem viable, the band is loosened a trifle and the case followed, in the hope that further treatment will not be needed and with the feeling that a collateral circulation is being developed

If the pulsation in the sac is not controlled, some other procedure to cure the aneurysm is required

Surgical Treatment of Popliteal Aneurysm—Undoubtedly the routine method used should be Matas's *Obliterative Aneurysmorrhaphy*, but for those skilled in the treatment of aneurysm, *Restorative* and *Reconstructive Aneurysmorrhaphy* are available and are occasionally useful

Obliterative Aneurysmorrhaphy—This operation, already described under Subclavian Aneurysm, is pictured in Figure 23 The afferent and efferent vessels having been brought under control, preferably by elevation upon a piece of rubber

of the vascular malformations and arteriovenous fistulas have the appearance of innocent superficial swellings, others take the form of large superficial vessels much like varicose veins, and yet others are pulsating masses, which call attention to themselves by a thrill and audible bruit, and are evidently dilated by receiving an arterial stream. At first sight, the relation between these various types is not clear, but basically all such as are not of traumatic origin belong to one family; that is, they are failures of development in the common capillary plexus of the embryo from which the arteries and veins are evolved. All are apt to be called angiomas, or hemangiomas, with the implication that they grow, and actually there are tumors, some of them malignant, which originate in vascular endothelium. However, the arteriovenous malformations rarely grow in the sense that their parts multiply. They enlarge because they swell as a result of dilation of the blood spaces or vessels of which they are formed. The simplest of them is a swelling composed of undifferentiated spaces, neither arterial or venous. Such is the "naevus", which may be "capillary" or "cavernous" and which may have the purplish covering of the port-wine stain, a coat of normal skin, or a mixture of both. By contrast, the arteriovenous fistulas are serious and occasionally terrifying lesions. The great arteries and veins are formed, yet retain connections from one to the other, so that at one or many places blood pours from artery into vein, causing obvious dilatation and carrying in its train some very remarkable changes, both in the part involved and in the circulation in general. In other words, it makes a great deal of difference whether the malformation occurs in the vascular bed of the skin and subcutaneous tissue or in the great vessels serving a limb.

Both the capillary and cavernous types of malformation are supplied with arterial blood but so indirectly and with so little force that they never pulsate—the tissue merely resembles a very vascular sponge with smaller or larger meshes. Another type has a more direct connection, usually by a series of tiny vessels, but there need be no actual pulsation in the

together by mattress sutures The controlling tubing at either end of the aneurysm can be loosened from time to time to see if hemostasis is satisfactory

Restorative Endoaneurysmorrhaphy—This operation is only possible in saccular aneurysms, especially when the original arterial lumen is well preserved, so that even if the lateral opening is large, the original channel is present as a groove The blood supply being controlled by tubing or rubber covered artery clamps and the sac laid widely open, the slit or gap in the side of the artery is closed with a continuous stitch of fine oiled silk reinforced with a number of individual silk or chromicized catgut stitches The current is then allowed to resume its natural course It must of course be determined that the efferent artery is not thrombosed

Reconstructive Endoaneurysmorrhaphy—This is the aneurysmal expert's dream which is almost never realized There is seldom any real need of the operation, which in any case is only possible of performance when there is present, from one end of the aneurysm to the other, a straight posterior lane of normal intima The walls of the aneurysm must then be sewed together in such a way as to leave a posterior channel Matas recommends that this channel be constructed over a piece of rubber tubing which is extracted before the long row of stitches, carefully placed, is finally tied

Should syphilis become rare or extinct, the arterial aneurysms of the lower limbs will be practically the only ones encountered Even today, many a surgeon goes through life without seeing any of them Probably the arteriovenous sorts are better worth studying

Abnormal Arteriovenous Communications

There is here presented a brief account of such unnatural arteriovenous connections, both congenital and traumatic, as are likely to appear upon the extremities, with the idea of sorting out the different varieties and distinguishing these uncommon and often serious lesions from the more familiar and harmless states which in some degree they resemble Many

of the vascular malformations and arteriovenous fistulas have the appearance of innocent superficial swellings, others take the form of large superficial vessels much like varicose veins, and yet others are pulsating masses, which call attention to themselves by a thrill and audible bruit, and are evidently dilated by receiving an arterial stream. At first sight, the relation between these various types is not clear, but basically all such as are not of traumatic origin belong to one family; that is, they are failures of development in the common capillary plexus of the embryo from which the arteries and veins are evolved. All are apt to be called angiomas, or hemangiomas, with the implication that they grow, and actually there are tumors, some of them malignant, which originate in vascular endothelium. However, the arteriovenous malformations rarely grow in the sense that their parts multiply. They enlarge because they swell as a result of dilation of the blood spaces or vessels of which they are formed. The simplest of them is a swelling composed of undifferentiated spaces, neither arterial or venous. Such is the "naevus", which may be "capillary" or "cavernous" and which may have the purplish covering of the port-wine stain, a coat of normal skin, or a mixture of both. By contrast, the arteriovenous fistulas are serious and occasionally terrifying lesions. The great arteries and veins are formed, yet retain connections from one to the other, so that at one or many places blood pours from artery into vein, causing obvious dilatation and carrying in its train some very remarkable changes, both in the part involved and in the circulation in general. In other words, it makes a great deal of difference whether the malformation occurs in the vascular bed of the skin and subcutaneous tissue or in the great vessels serving a limb.

Both the capillary and cavernous types of malformation are supplied with arterial blood but so indirectly and with so little force that they never pulsate—the tissue merely resembles a very vascular sponge with smaller or larger meshes. Another type has a more direct connection, usually by a series of tiny vessels, but there need be no actual pulsation in the

receiving veins nor any bruit or thrill The connection may indeed be so insignificant that the blood of the prominent veins which mark the lesion is merely given an arterial tint and a rather high oxygen content Occasionally it happens that one of these relatively quiet forms is injured or undergoes some other unexpected change by which the afferent artery is able to pour a large stream *directly* into it Then the original angioma and the efferent veins dilate and pulsate, forming what has often been called a cirsoid aneurysm This is most apt to occur upon the scalp

The arteriovenous aneurysm or fistula is especially common at the root of the neck or of a limb, because in these situations great arteries and veins lie rather superficially in close contact and are held together in a fibrous sheath. If they are malformed, that is, not fully differentiated from one another, there may be multiple fistulas between them, a series of small channels making connections over a considerable distance, as, for instance, for the length of a thigh, or even through a good part of the arm and forearm A fistula has usually been defined as a direct opening or simple narrow passage, whereas an aneurysm is held to be a sacculation interposed between the artery and vein The cirsoid aneurysm then is merely one which presents a varicose appearance Such distinctions, even if valid, are of no basic consequence

The traumatic arteriovenous fistula differs in having only one connection (unless one artery communicates with two veins) and that usually a relatively large one such as may be made by flying glass or steel, by the stab of a narrow blade or a bullet wound Some of the aneurysms of the ancients were probably of this kind and must have been made by spears or arrows However, pure arterial aneurysms must also have occurred even though syphilis was apparently unknown before the end of the fifteenth century Thus the operation of Antyllus—ligation of the afferent and efferent vessels and removal of the sac—which has been handed down as a classical procedure, may have been used for lesions of either sort A common site for the acquired fistula in these days is the region



Abnormal Arteriovenous Communications *A.* Cavernous angioma Treatment by use of carbon-dioxide snow, followed by partial excision (courtesy of Dr D W MacCollum, Children's Hospital, Boston). *B* Extensive cavernous hemangioma in process of treatment by carbon-dioxide snow. *C.* Prominent but local cavernous hemangioma, treated by excision (courtesy of Dr Robert E Gross Children's Hospital, Boston) *D.* A seemingly innocent capillary nevus The infra-red photograph on the right shows how large were the veins with which it was connected (courtesy of Dr. George D. Cutler, Children's Hospital, Boston)

PLATE X



ABNORMAL ARTERIOVENOUS COMMUNICATION A huge vein fed by very small arteries—associated with a capillary nevus of front of thigh Treated by excision, and by the subsequent injection of sclerosing solutions Note subcutaneous hemorrhage in the postoperative photograph G C 15913

PLATE XI



ARTERIOVENOUS FISTULA, CONGENITAL J.J.M., 51848, a man, aged nineteen Infra red photograph before operation See arteriogram, Plate XIII, A and B

PLATE XII

A

Radial artery

Anterior interosseous artery

Ulnar recurrent artery

Ulnar artery

B

Radial artery

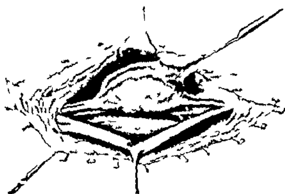
Anterior interosseous artery

Ulnar recurrent artery

Ulnar artery

M Codd

J F M, 51518 Same patient as shown in Plate XII Sketches after arteriograms made following the first operation in which only the proximal of many fistulas was divided *A* First exposure Proximal connections with contorted mass of veins on back of arm *B* Second exposure (3 seconds later) The radial and ulnar arteries free from fistulas are not filled by the thorotrast The fistulas arise chiefly from the ulnar recurrent and anterior interosseous arteries The flow follows the course of least resistance, that is, through the various fistulas





C An Arteriovenous Aneurysm of the Brachial Artery—following a wound by a fragment of metal A case treated and published by Drs Reid and McGuire who have kindly permitted a reproduction of their illustration (Courtesy of Annals of Surgery, 108 643 Oct, 1938)

PLATE XIII

behind or above the clavicle, less frequently the groin, and the bullet is perhaps the most common cause. The nature of the great mass of pulsating veins which soon develops and of the changes in the heart which so often follow will be explained in a subsequent account of the lesion.

Hemangioma: Capillary and Cavernous

The familiar and picturesque deep purple birthmark upon the face or scalp, which so accurately occupies the field of one of the trifacial branches is a rather pure hemangioma, very superficial and capillary or deep and cavernous as the case may be. Not all congenital malformations are so clean-cut. Some subcutaneous swellings are unclassifiable, being disorders of the subcutaneous structures in general, lymphatic as well as arteriovenous, as is told in the following chapter. The hemangiomas as a rule are compressible, that is, much of their content can be expelled by pressure and returns on release. No vessels are seen entering or leaving them, nor do they pulsate. Such as these occasionally appear *on the limbs*. They are usually raised above the surface and sharply marked off from the normal surrounding skin. Their covering may be wholly of the purple capillary sort or of a reasonably normal skin. But there is always some admixture of port-wine stain in the form of smaller or larger patches. Multiple lesions are sometimes seen. Several of these malformations, all of the cavernous type, with a more or less capillary surface are shown in Plate X. These are taken from the records of the Children's Hospital in Boston, to the staff of which the writer is much indebted.

Treatment is usually surgical excision. Whereas on the face or scalp the large hemangioma must often be treated by the application of carbon-dioxide snow, by multiple punctures with the endothermy needle or perhaps by radium or the X ray, it is seldom that those of the limbs can not completely be excised. They may be fed by several large vessels, but since the operation can be kept in the field of normal tissue about them, the control of their blood supply is not too difficult. The

muscular aponeurosis is apt to be missing, so that the base of the angioma must be dissected from the underlying muscle The lesions shown in Plate X, have been treated by various methods or combinations of methods as is told in the legends

Tumor of the Cutaneous Glomus Glomangioma—This rather rare tumor is especially worth having in mind because it has the peculiar quality of being extremely sensitive and painful Long known as "subcutaneous painful tubercle" it was first shown by Masson (1924) to represent an abnormal development, that is, a hemangioma, of the cutaneous glomus, the widely distributed tiny neuro vascular organ which governs the rapid vasomotor reactions of the extremities By its means, the arteriovenous flushing passages in the deep skin of the hands and feet are opened (Chapter I) raising the temperature of the surface In the upper limb, the glomus tumor is most often found upon the fingers, the thenar and hypothenar eminences, and especially beneath the nails In the lower, it is more erratically distributed Rarely it is seen upon the body

Attention is called to the glomangioma by a sensitive spot which causes the individual to protect it against all contacts, whether by friction or pressure Pain of a knife like, radiating sort, is also excited by cold and is even spontaneous, but for all this sensitiveness Bailey remarks that most patients harbor the tumor for many years before seeking relief Over it, the skin may be normal in color and without elevation, or may be bluish and elevated Under the nail it makes a purplish spot Actually, the glomangioma is so very vascular that, when exposed, it must display a color somewhere between red and blue Its size is small, rarely more than a centimeter (one fourth to one half inch) in diameter and it does *not* grow, having no malignant tendency whatever

Pathologically, the tumor is distinguished by showing certain epithelial glomus cells, a considerable admixture of fine nerve fibers and structures reminiscent of the contorted vessels of the typical glomus

A very interesting feature, first noticed by Barre, for

which watch should be kept, is the association of the glomangioma with a chronic sort of vasomotor change in the direction of heat and flushing (or coldness and cyanosis) of the extremity.

Treatment is surgical excision, which offers no especial difficulty and is entirely curative. A nerve block with procaine should be used wherever possible, rather than local infiltration.

Congenital Arteriovenous Fistula or Aneurysm

The appearance of this lesion varies enormously according to the nature and extent of the connection between artery and vein. There will perhaps have been noticed from birth a group of veins upon some part of a limb, a patch easily distinguished from the area about it and without visible superficial afferent or efferent vessels. Often the skin over some parts or all of the patch will be more or less abnormal in appearance, a little purplish or brownish. Sometimes a long dilated vein or plexus of veins will pass down an extremity. Along its course the skin will show blotches or spots of discoloration and appear slightly pitted or irregularly bossed. Usually such a lesion will take roughly the form of a broad band or occupy part of a foot or hand following in an erratic way one or more dermatomes.* In other words, although large veins may be present, there is apt also to be a suggestion of the port-wine stain, the capillary nevus.

A mass of veins or a single vein of this kind does not pulsate. Indeed, if its anatomical situation is such as to correspond to the familiar saphenous varicosity, it may be mistaken for varix. There is no swelling of the limb but there will occasionally be lengthening of some bone. The writer has seen a rather local lesion of this sort, chiefly confined to the outer side of the lower leg, external malleolus, and dorsum of the

* The dermatomes upon the extremities correspond to the sensory zones upon the body which take their origin from the spinal segments. Those of the arms and legs are elongated and have no representation upon the body. Their number corresponds to that of the nerve roots which furnish the nerve supply, respectively, for the upper and lower limbs.

foot, which led to lengthening of the leg at one period of the child's growth, though subsequently the two limbs became equalized The skin over such an angioma will perhaps feel warm to the touch as compared with other parts It is impossible, without a study of the blood drawn from the distended vein, to determine the size of the arterial fistula In the absence of a bruit, the arterial connection can only be very indirect, yet it may be sufficient to cause considerable difficulty in controlling bleeding both at the time of excision, if a surgical operation is used, or afterwards, as the following case shows

G C, a young man twenty one years of age, had noticed all his life a dark patch of skin upon his left thigh and a large vein which seemingly emerged from the muscle on the upper, external surface of the thigh near the gluteal fold and ran down nearly to the ankle By the Trendelenburg test, this vein lacked valves It was in fact varicose The lower limbs were otherwise symmetrical No pulsation could be felt and, apparently, before surgical treatment was undertaken no bruit was searched for, yet subsequently a faint and utterly nonrhythmic, bubbling or clicking noise was heard through the stethoscope over the discolored area of skin above the knee Blood from the vein was unfortunately not examined

Through a long incision in the thigh and another in the lower leg the greater part of the vein was excised The vascularity of the tissues was noted and at one or two points, notably where the vessel emerged from an opening in the fascia lata, bleeding was difficult to control Throughout its course in the thigh the vein gave off a series of "infinitely small branches apparently connecting with the naevus" which lay principally median to it Healing was complicated by an accumulation of blood clot in several parts of the wound, so that the patient was unable to leave the hospital for six weeks

Ten months later G C returned for the treatment of an ulcer and shallow sinus at the upper end of the wound in the thigh Serious bleeding had occurred from this point on several occasions Upon excision of this area a plexus of large

veins was found, lying partly upon and partly beneath the fascia lata. Painstaking removal of the plexus was followed by permanent healing. Three years later, various dilated tortuous veins in the lower leg were successfully treated by the injection of sclerosing solutions and at the present time, sixteen years after the original operation, the leg gives no further trouble. It looks very much as it did in the photographs taken immediately after the first operation (Plate XI).

Cases much like the above have many times been described. In one of those recently published by De Takats, who has very kindly permitted me to reproduce several of his sketches, an additional and very troublesome feature was the presence in the birthmark of very delicate multiple capillary angiomas

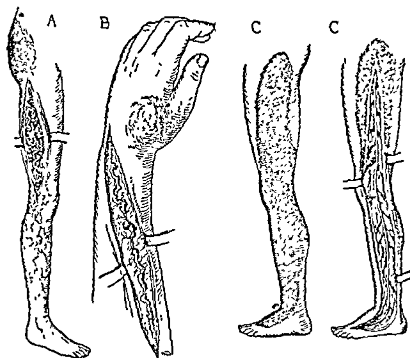


FIGURE 24. ABNORMAL ARTERIOVENOUS COMMUNICATIONS. (By courtesy of Dr de Takats: Published in *Surgery, Gynecology and Obstetrics*, 55·227-237 (Aug) 1932 Figures 6, 8 and 12) These bring out the associated capillary nevi and, in one case, *C*, peculiar spots consisting of delicate collections of vessels which bleed easily.

which bled readily By such lesions the nutrition of the extremity is in no way harmed, the circulation in general is not affected, and only if the angioma chances to increase the blood supply of the growing end of a bone or bones will any effect upon development be observed, that is, the extremity may be overgrown A very different sort of disorder is caused by injury to the hemangioma Then dangerous external bleeding may occur, or, as already explained, if the connection of an efferent artery with the lesion is suddenly widened, a bruit or thrill or even a palpable pulsation may develop The distant results of such a change require separate description

The Physiological Changes Consequent upon Large Arteriovenous Fistulas or Aneurysms

These changes have long excited the curiosity of those who have been obliged to deal with such disorders For a historical account of the many fundamental observations upon this subject, Holman's monograph should be consulted In this country, Matas, Halsted, Reid, and Holman, in particular, have made significant contributions both to the study of the lesion as well as to its treatment Most of the observed clinical findings have been reproduced experimentally by Holman It appears that the secondary effects, namely, dilatation of the afferent artery and of the heart, are proportional to the size of the fistula Considerable time is required for their development and this again is related to the size of the unnatural connection Moreover, the nearer the heart the fistula is situated the more rapid and extensive will be the secondary changes With such modifying circumstances in mind, the following may be considered to represent the usual result of the serious arteriovenous communication The explanations given are the result of much experiment and observation, but need not necessarily be considered correct

General Effects upon the Heart and Circulation—A large fistula calls for unnaturally rapid and vigorous contractions of the heart, increases the cardiac output, causes dilatation and perhaps hypertrophy of the heart, and may in time lead

to decompensation. When arterial blood pours from a large artery into a vein, the circulation in the limb, or neck, or organ, as the case may be, is short-circuited. An easy path is made, by which the normal, resistant vascular bed of small vessels and capillaries is avoided. As between the distal capillary bed and the fistula, the stream therefore chooses the fistula. The sudden flooding of the capacious venous system which necessarily occurs has the effect of a massive hemorrhage. The arterial pressure falls and, in consequence, the heart beats faster. Venous pressure is elevated. Sufficient blood now being supplied to the right side of the heart, more blood is put out per beat and the arterial pressure rises. In this way the systolic pressure may gradually be restored to a normal figure but the diastolic pressure remains low, the pulse pressure being increased much as an aortic regurgitation (Lewis and Drury). The tendency of a fistula, then, is first to lower and then raise blood pressure, raise the pulse pressure and increase the pulse rate.

To compensate for the loss of blood on the arterial side, which results from flooding the veins, an increase in blood volume occurs,* and the cardiac output becomes greater. Yet the fistula continues to carry much of the larger volume into the shorter circuit and so, to accommodate the increased bulk of blood, the heart and blood vessels dilate. Only the large fistulas induce this change and especially those not too far out toward the periphery. A fistula between the external iliac artery and vein is more potent than one in Hunter's canal.

The converse of these changes has repeatedly been demonstrated. If the fistula is temporarily closed by pressure, the heart rate is slowed, the blood-pressure rises. When the fistula is permanently closed, the blood-volume is soon reduced, the blood-pressure, temporarily elevated, again falls, and the size of the heart is restored to normal.

* This Holman maintains on what appear to be satisfactory experimental and clinical grounds. Reid is inclined to disagree. However, no one disputes the fact that the heart rate is increased, that the heart puts out more blood with each beat, and that the heart tends to dilate.

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The Progress of an Arteriovenous Fistula.—A small fistula is very occasionally closed by thrombosis and organization, one of modest size often remains stationary, but a large one tends to become larger. As the afferent artery and efferent veins dilate, there is a natural and purely mechanical tendency to widening of the fistulous opening. As the opening widens, peripheral resistance diminishes, more blood passes through it, the *total* volume of blood increases, the heart and proximal arteries dilate further, and so the fistula enlarges still more, a true vicious circle. If, however, at any time, pressure in the *receiving* veins rises enough to equal the peripheral resistance beyond the false passage, dilatation of the short-circuiting fistula will cease, and the vicious circle will be broken. By this same equalization, the dilatation of the heart is brought to an end, and ultimate degeneration of that organ may be prevented. Actually, in a young individual, the increased blood volume and elevated cardiac output are compatible, even over many years, with a heart of normal size and behavior.

Summary of the Effects of an Arteriovenous Fistula

1. The *effects* are greater according as the fistula is (*a*) larger and (*b*) nearer the heart.
2. The *arterial* blood pressure falls.
3. The pulse rate is increased.
4. The venous pressure rises (between fistula and heart and perhaps elsewhere).
5. The blood volume is increased.
6. The cardiac output is increased
7. The heart becomes dilated (and perhaps hypertrophied).
8. The afferent *artery* becomes dilated and weakened.
9. The efferent *veins* become dilated and strengthened.
10. The collateral circulation about the fistula to peripheral parts develops.
11. The limb may become overgrown, but its *terminal* part may become ill-nourished.
12. All these effects are *reversible* If the afferent and efferent vessels are divided and the *fistula* removed, the peripheral part is well nourished by the collateral circulation. Indeed nutrition of the peripheral part will be improved.

Local Effects upon the Blood Vessels and the Peripheral Parts—The tendency of a fistula is toward the "venafication" of the proximal artery, toward "arterialization" of the receiving vein and toward the establishment of an abundant collateral circulation Reid points out that under the influence of the lowered resistance of the fistula and the lack of recoil against the arterial wall, the latter seems to undergo a sort of degeneration Its muscular and elastic fibers deteriorate and it soon dilates Possibly, as Holman suggests, the greatly increased blood mass of the shortened circuit enters into this dilatation In any case, the process is often carried well back toward the heart and very rarely occurs distal to the fistula It may be so serious that after excision or repair of the fistula, the afferent artery becomes an aneurysm or actually ruptures The receiving vein, by contrast, not only dilates but hypertrophies All coats are thickened and strengthened, the elastic material being greatly increased This results in one of the most conspicuous features of the large fistula, the great mass of dilated, pulsating veins

The effect of this local change upon the peripheral part of the limb, in the region of and for some distance beyond the false opening, is to enlarge it and to make its surface unduly warm Peripheral to this swollen, hot area, the extremity is left cool and sometimes actually ill nourished, so that local necrosis and ulceration may occur Strangely enough, however, a collateral circulation of considerable importance is developed, as a result, seemingly, of the very low pressure in the vessels near the fistula Possibly the oxygen want established in the peripheral tissues hastens the peripheral vasodilatation In any case, the collateral channels, if established in the region of a growing epiphyseal line, sometimes occasion overgrowth of a long bone Moreover, if the fistula, together with its afferent and efferent vessels, is excised, the collateral circulation can be relied upon to nourish the extremity However, if the afferent artery *alone* is divided, the collateral circulation carries all its blood into the fistula (easy retrograde pathways), and peripheral gangrene results

greater in circumference than the right. Its upper part was hot to the touch. The hands were alike. Great veins could be seen on the ventral surface of the whole forearm, but there was an especially prominent mass of them just below the back of the elbow. There was felt, over the region of the elbow and forearm, a systolic thrill, and a continuous bruit was audible through the stethoscope. Thrill and bruit extended along the course of the brachial vessels into the axilla, ending at a point above the clavicle. The radial pulses were alike and seemed of similar force. Firm pressure on the lower brachial, just above or in the antecubital fossa, obliterated both bruit and thrill. Pressure over the great mass of pulsating veins upon the dorsal surface of the upper forearm nearly but not quite accomplished the same thing.

The following special observations were made by Dr. C. S. *Burwell* and his associates of the Medical Staff of the Peter Bent Brigham Hospital.

1. Pulse rate with fistula open, 84
   Pulse rate with fistula closed, 71.

2 Arterial Pressure	*Right Arm*	*Right Leg*	*Left Leg*
Fistula open	102/57	123/83	128/76
Fistula closed	117/78	142/100	142/87

3. Venous Pressure* (in mm. of water)	*Right Arm*	*Left Elbow*	*Dorsal Aspect of Left Forearm*
	65 mm.	145 mm.	265 mm.

4. Circulation Time	*Right Arm Vein to Tongue*	*Left Arm Vein (just above antecubital space)*
	22.5 seconds	12 5 seconds

5. Oxygen Content of Venous Blood in Volumes per cent. (Oxygen capacity 18.2)	*Right Arm*	*Left Arm*
	14.5	16-17 in forearm 14 3 at wrist

*Elevation of the venous pressure, in this case at least, is apparently a local one. It is most marked in the vicinity of the fistula and diminishes progressively as it is examined nearer the heart. However, at every point studied between the fistula and the heart, the venous pressure was higher than in other parts of the body.

The Congenital Arteriovenous Fistula Case Report

It has already been explained that there is no valid distinction between arteriovenous fistula and aneurysm, but there is a rather consistent difference between congenital and traumatic communications When in the course of the differentiation of artery from vein in the common capillary bed of the embryo, connections are left uniting the two sorts of vessels, these are apt to be multiple, to occur even over a considerable distance Especially upon a limb, there may be a whole series of small connections between the principal artery, or one of its larger divisions, and its companion vein A traumatic fistula, on the other hand, unites the paired vessels at some one spot, in the axilla, at the elbow, the groin, the popliteal space, or some intermediate point And because a considerable amount of blood escapes into the tissues where the bullet or sliver passed through the two vessels, the resulting hematoma is apt to become organized and to be hollowed out into a false aneurysm connecting with both artery and vein and finally lined with endothelium Thus, most congenital communications resemble fistulas while the traumatic ones are either direct openings or arteriovenous aneurysms In such a situation as the neck, the two sorts of lesions resemble each other, but in a limb the congenital lesion is apt to present a characteristic appearance The following case report gives an account of most of the features of a congenital multiple fistula in an extremity and of some of the difficulties met in treating a condition of this sort (See Plates XII and XIII, A and B)

J J M, a husky nineteen year old youth, had noticed nothing remarkable about his left arm until, at the age of ten, he fell out of a swing Then the arm began to swell but never lengthened disproportionately Dilated veins appeared about the elbow and upper forearm, especially upon its dorsal surface and after a time were seen to pulsate The arm caused no pain or disability but because of the pulsation and palpable thrill the boy was induced to seek treatment.

Examination showed the left forearm to be seven cm

exposed. The brachial was larger than before and seemed very friable. Several small connections from artery to vein were found just above the bifurcation. Upon dividing these, the ulnar and the ulnar recurrent arteries were seen to have large connections with veins, but since distal division of these arteries and veins could not be performed from the antecubital side and since it was evident that distally placed fistulas would continue to open up as the proximal ones were obliterated, the wound was closed and further surgery was for the moment abandoned.

During the next two weeks the dilated brachial artery ruptured twice (others have noticed this accident) and was finally divided, together with the vein, half way from elbow to shoulder. The median nerve was accidentally severed and was reunited by suture.

The result, January, 1938, shows considerable improvement. The pulsation, thrill, and bruit in the forearm are gone. The hand is well nourished. However, the bruit can still be heard in the axilla. The median nerve has shown a reasonably complete regeneration.

This case illustrates the multiple character of the malformation, the dilatation and degeneration of the afferent artery, and the opening up of secondary peripheral connections as the proximal ones are closed. More especially it shows how remarkably efficient is the collateral circulation created in the presence of such a fistula. Ordinarily, division of the efferent artery results in gangrene of the limb (the blood carried by the collateral vessels flows back into the fistula). Here the proximal vein, as well, was divided (raising the pressure on the venous side and forcing blood to follow the collaterals) and successive division of several proximal fistulas probably helped to increase the resistance of the fistulous pathway and direct the stream into the peripheral parts.

Traumatic Arteriovenous Aneurysm and Fistula

In order to make clear the distinction between the various lesions which may result when an artery is pierced or rup-

6 Total blood volume 6770 c cm or 102 6 (c cm) per kilo (Blue dye method of Gibson and Evans) "which is well above normal even for a very muscular active man and is at a level seen in congestive heart failure"

7 The heart, by seven foot plate, was within the normal limits of size

Operation, June 4th, 1936 Under ether anesthesia and with compression of the upper arm by an Esmarch bandage, an incision was made exposing the brachial vessels in the lower arm and through the antecubital space In the upper part of the wound, for a distance of five to six cm, the brachial artery appeared twice its normal size It then became quite rapidly smaller, reaching its natural caliber some two to three cm above its division into radial and ulnar branches Almost four to five cm proximal to the point where the brachial narrowed, was an S shaped little vessel two to three mm in diameter and about one and a half cm in length which passed from artery to vein This was divided and tied with silk Then the operator, not at first realizing that this little vessel might be an important fistula, asked to have the Esmarch bandage loosened and when the current was let in, the venous pulsation, thrill, and bruit in the region of the elbow were found to have ceased!

However, an axillary thrill and bruit persisted and since these in turn were blotted out by compressing the brachial artery and vein in the wound, some connection distal to the division of the brachial at the elbow was judged to have persisted This idea was confirmed on the following day when the expansile pulsation, bruit, and thrill in the forearm returned

Two months later an arteriogram was made This showed that "the opaque material enters the region of the aneurysm by means of the dorsal and volar interosseous arteries and the ulnar recurrent artery, the radial artery does not appear to communicate except through the radial recurrent"

Second Operation, October 7th, 1936—The former scar was excised and the lower brachial artery and its bifurcation again

point and controlling its blood flow with a rubber-covered clamp or rubber tubing, or, if this is impracticable, by making digital pressure upon it, the clot and blood can often be evacuated from the region of injury and the artery either repaired by suture or ligated above and below the opening. Naturally, the decision whether or not to attack the vessel directly or to allow a hematoma to develop to its fullest extent is a difficult one. Provided means of transfusion are at hand, direct attack is usually the method of choice.

A pulsating hematoma which has actually come to resemble a purely arterial aneurysm will present the signs of aneurysm. That is, it will have an expansile pulsation and a *systolic* bruit and thrill, in contrast to the arteriovenous aneurysm or fistula in which these signs, though accented in systole are continued through diastole. The treatment of such a false aneurysm is that of aneurysm (*q.v.*).

Varieties of Arteriovenous Aneurysm and Fistula

It would be impossible to describe all or indeed many of the appearances presented by traumatic fistulas in various regions. Naturally most of the victims will be aware that they have been shot or stabbed, though a fine, flying sliver of glass or steel may perhaps fail to be noticed. Some bleeding from the injured artery is inevitable. External bleeding may occur or a large hematoma may form. However, a fistula is sometimes established so quietly that a continuous audible bruit and palpable thrill, appearing a few days later, will be the first signs of the seriousness of the injury. Occasionally, especially in wounds of the limbs, the aneurysm or fistula will make pressure on a near-by nerve, causing numbness or muscular paralysis.

The rapidity with which cardiac damage may take place after the first signs of fistula have been noticed, is vouched for by a case reported by Mason, a brief account of which is abstracted from his report.

A colored woman, aged thirty, was first seen half an hour after being stabbed below the left clavicle, close to the sternum.

tured it is necessary to consider several possible occurrences (1) the artery alone is injured, in which case a *Pulsating Hematoma, or False Aneurysm* may develop, (2) the artery and vein are injured in such a way that, following the organization of a hematoma, a connection is established between them by way of a sac, which becomes lined with endothelium—an *Arteriovenous Aneurysm*, or (3) the artery and vein are so injured that a direct connection is made between them—an *Arteriovenous Fistula* For purposes of study and treatment, it has already been stated the arteriovenous aneurysm and fistula are alike However, the pulsating hematoma is different and requires a separate description

Pulsating Hematoma, or False Aneurysm —This is a very rare lesion and the reason for this is easy to see When a large artery is pierced, its companion vein is usually injured as well, so that if the individual survives and if the vessels are not at once ligated or repaired, an arteriovenous connection is made Suppose, however, that an artery alone is injured The opening is made beneath a heavy layer of fascia If it is not so directly connected with the outside world that the individual bleeds to death, a hematoma is rapidly formed beneath the aponeurosis Bleeding continues until the pressure within the hematoma equals the arterial pressure If the hematoma is then left to itself, it will tend to become so tense as to block the blood supply to the distal portion of the limb (in the case of the femoral, popliteal, axillary, or brachial, for instance) and gangrene will ensue This is all the more likely to happen because the rapid loss of blood will have caused a decided fall of blood pressure However, should the limb survive, the mass of extravasated blood will soon become organized at the periphery, its center remaining liquid Only through this very rare combination of circumstances can a pulsating hematoma form, but actually it is seldom given an opportunity For if an individual has been stabbed or shot in such a way that a large artery appears to have been injured and if a hematoma is rapidly enlarging, most surgeons will endeavor to ligate the vessel By approaching it at a proximal

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ard treatment for arteriovenous fistula. Matas has successfully restored the continuity of the subclavian by closing the *rent in the artery* through the opened vein, but most surgeons feel more sure of a cure in making the quadruple division and *excising the fistula*-connected vessels. Mason gives to everyone who contemplates attacking a subclavian arteriovenous fistula the advice to begin by reading Halsted's essay on "Ligation of the Left Subclavian Artery in its first Portion". The difficulty of access is such that one should always resect the clavicle and even the adjacent upper and lateral portion of the manubrium sterni. The clavicle is missed less than one would suppose and if resected subperiosteally will often regenerate.

Subclavian fistulas bring up most of the difficulties inherent in treating all traumatic arteriovenous communications. If the operation is performed too soon, a collateral circulation may not have become established. If performed too late, the heart will have received permanent damage or the patient will have deteriorated beyond aid. If performed early, the vessels will have barely recovered from the injury and will be difficult to handle. If performed late, they will have established so many unexpected connections that quadrilateral ligation will fail to still the current in the region of the fistula. This last difficulty was demonstrated to the writer by Elkin in an operation upon a subclavian fistula of sixteen years' duration. Incidentally, this case, by contrast with Mason's, though it had caused the heart to dilate, had led to no cardiac decompensation. After three hours' work and when every visible connection with the great dilated vessels which constituted the arteriovenous aneurysm had been divided, arterial blood still passed abundantly through the fistula. Any further approach turned out to be so very bloody that recourse was had to infolding the remaining vessels with a series of heavy stitches. Reid has faced this situation in a similar way, and on another occasion, when all the vessels connected with the fistula could not be severed, has twisted the divided proximal vein until it was not only obliterated but completely closed the opening.

To summarize the subject of the subclavian fistula: it may

Her heart was known, by previous examination, to have been normal and [illegible]
succeeding
region, and a bruit could be heard locally and in the neck Thrill and bruit, which were continuous, increased in violence and range during the two following days

Sixteen days after the injury, the signs persisted, but the heart appeared undamaged The same was true after thirty three days

About forty-five days after the injury, the patient was discovered confined to bed by circulatory embarrassment and at *the end of nearly ten weeks was in a serious condition* The heart was enormously dilated, a pleural effusion had formed on the left, the feet and legs were edematous, the liver enlarged, the abdomen distended and dropsical Dyspnea and cough were present *For all this*, there was as yet *no external* swelling over the fistula.

Two operations, under a local anesthetic, were required before the fistula could be isolated by quadruple ligation and excised, so that it was over three months after the wound had been received before the strain of the great fistulous leak from the subclavian artery could be taken off the heart As a result, the heart regained its normal size and all the other signs of decompensation cleared up, but an aortic thrill and bruit remained as well as a double mitral murmur Mason noted the occurrence of Branham's sign when he drew tight the ligature on the subclavian artery, that is, the pulse rate fell at once (from 124 to 104) Had this difficult operation not been so skillfully and bloodlessly performed the patient would soon have died

Subclavian Fistula —The case just quoted will serve as an introduction to this subject It has been duplicated by others, though never more dramatically There is a general agreement that division of the artery and vein proximally and distally, that is, quadruple ligation, completely isolating the fistula, is compatible with the life of the limb supplied by the injured great vessels This may then be regarded as the stand-

tive. The radial pulse was not altered. All vessels were found to have opened into a common sac.

Here the early operation was forced upon the attendant surgeons, yet, perhaps because the young patient's vessels were so elastic, quadruple ligation and excision were followed by no disabilities.

It is hardly necessary to describe other arteriovenous fistulas of the upper limb. Such a lesion may even occur on a finger. Elkin illustrates one of this sort. The treatment of such conditions is exactly that of the larger vessels.

Femoral Fistula.—In general, the fistulas of the lower extremity are less well borne and more prone to cause cardiac dilatation and decompensation than those of the upper. Those of the femoral, because of its size, are particularly serious. The limb is apt to become edematous and cyanotic, and sores upon the lower leg may develop. Though the lesion is easily accessible, the many vessels centering on the region of the groin make it especially complicated. The inferior epigastric and deep circumflex iliac artery and vein, the great saphenous vein, the profunda femoris artery and vein, all join the superficial femoral almost at one spot. Thus a fistula near the inguinal ligament may be impossible of excision. Even after dividing the proximal and distal vessels, the fistula-connected artery and vein will perhaps continue to exhibit the familiar thrill. It may then be possible only to transfix and plicate the actual region of the false passage with many stitches. As in treating fistulas of other great vessels, it is usually necessary, as Reid tells, to ligate with tape, rather than even the largest and heaviest silk. Halsted's metal bands may be useful on occasion, especially when the proximal artery is dilated and friable.

Popliteal Fistula.—This, like the true arterial aneurysm, is especially apt to interfere seriously with the nutrition of the foot. Edema and cyanosis will usually be considerable. A fistula in the lower femoral or upper popliteal region is easier to deal with than one in the lower angle of the popliteal space. For here the popliteal artery, having given off the anterior

come on quietly or with a hematoma, it may lead rapidly to cardiac decompensation or be well borne by the patient, it may and usually does create a permanent mass of pulsating veins above the clavicle At the risk of some cardiac damage, it had better not be attacked surgically for several months, to permit the establishment of a collateral circulation It should be approached by an ample incision and by removal of the clavicle (and perhaps a portion of the sternum) It should be isolated and if possible excised after quadrilateral division

Fistulas of the Upper Limb—The same general principles as have been discussed for the management of fistulas nearer the heart apply to these, but the indications for treatment may be somewhat different By the kindness of Reid and McGuire, the writer is permitted to reproduce their illustrations of a brachial fistula, successfully treated by a very early operation

The patient, No 81335, Cincinnati General Hospital, a boy, sixteen years of age, had been struck in the right arm, just above the elbow, by a fragment of exploding pistol shell Free external bleeding was followed by marked swelling which subsided after three days At this time, a pulsating tumor appeared, and the usual continuous thrill and bruit were noted There also developed a weakness in gripping the hand and a tingling sensation in the hand (pressure upon the median nerve) The tumor rapidly and painfully increased in size

By the twenty third day after the accident, the median nerve palsy had become so pronounced, the causalgia so serious, and the aneurysmal covering so thin, that a surgical operation was imperative The circulation time in the right arm proximal to the fistula was *14 0* seconds and compared to *21 3* seconds in the left The heart was normal by every test Blood pressure was 120/60 in both arms, a suggestively low diastolic pressure On occluding the aneurysm, the pulse rate did not change but the diastolic pressure rose thirty mm. Hg

Operation disclosed the characteristic picture shown in Plate XIIIC The afferent vessels had not yet become dilated, and excision after quadruple division (or rather sextuple, since the two venae comites were involved) was cura

of blood in circulation may require the withdrawal of some of it after the fistula has been excised.

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tibial, divides into the peroneal and posterior tibial branches It is easy, then, to understand that when these are connected with the fistula or with the aneurysmal sac, division of all afferent and efferent vessels is almost impossible It is not as if one could control the arterial supply merely by compressing the main vessel proximal to the fistula. Cut off this vessel, and the distal branches at once pour in a retrograde current It may thus be necessary, after all accessible arteries and veins have been ligated, to plicate the remains of the sac, or the vessels joined by the fistula, with heavy silk

COMMENT ON THE TREATMENT OF TRAUMATIC ARTERIOVENOUS ANEURYSM AND FISTULA

In planning and executing a surgical operation, the following considerations must be kept in mind

1 The collateral circulation must have time to develop, but if more than a few months are allowed to pass, the local connections of the fistula may become so numerous and complicated that quadruple ligation is insufficient to effect a cure Then, special means may be required *to obliterate the fistula and its connecting vessels*

2 If treatment is delayed, serious damage may be done to the heart, which, therefore, must be very carefully watched

3 If the heart is so damaged that it can not be expected to withstand a radical operation, division of the proximal vein will lighten its load and perhaps permit it to recover

4 Ample exposure is absolutely essential

5 In exposing the fistula, the afferent artery should be reached and isolated as soon as possible and should then be controlled by the use of a piece of soft rubber tubing

6 The afferent artery should never alone be divided If division of the afferent vessel seems imperative, the afferent vein or veins should also be ligated

7 The afferent artery is often exceedingly friable and requires great care in ligation It may subsequently undergo aneurysmal dilatation *or actually rupture*

8 Since loss of blood at the operating table may be excessive, a sterile system of suction should be used to conduct blood from the wound into a receiving bottle containing sodium citrate It can then readily be infused into a vein while the operation proceeds

9 If very little blood has been lost during the operation, the excess

CHAPTER VIII

LYMPHANGIOMA ELEPHANTIASIS LYMPHEDEMA

The lymphatic system is an independent anatomical unit, having the function of absorbing all such tissue fluids as the blood capillaries are unable to accept and, in addition, foreign particulate matter. These fluids, filtered through intervening groups of lymph nodes, it forwards to the superior vena cava. In many ways, it resembles the venous system, with which it makes only two connections. Of these, the principal one is at the point where the left jugular vein joins the subclavian. Here the thoracic duct, carrying lymph from the lower limbs and lower half of the trunk, chyle from the lacteals, in fact, the lymphatic drainage from all of the body except the right upper quadrant, enters by a broad delta. The small, short duct on the right, receiving only the lymph from the right arm, right pectoral region, and right side of the neck and head, enters the right subclavian vein at a corresponding point. The lymph vessels, down to those of the very smallest size, are furnished at frequent intervals with valves, so that lymph is forced through them by muscular pressure from without, much as is the case with the veins. Indeed, without some movement, lymph tends to accumulate in the limbs and its flow is decidedly quickened by active exercise.

The ultimate lymph spaces, in the form of an epithelial-lined network, permeate the skin and subcutaneous tissues everywhere, a closed system of capillaries soon gathered together into somewhat larger but still hardly visible vessels which take very much the same course as the veins. The long lymphatics appear to run in several planes, (1) a very superficial one, in the true skin, where they are easily seen when infected and inflamed, as in the case of acute tubular lym-

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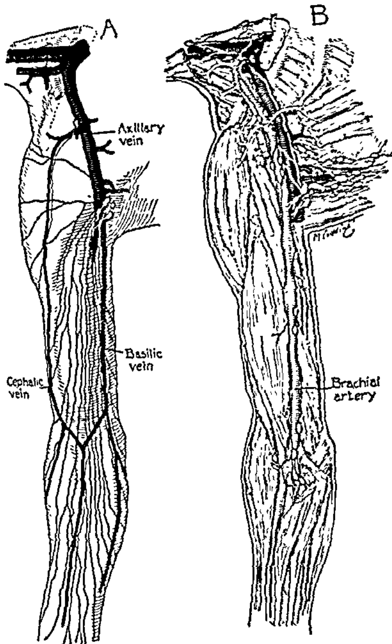


FIGURE 26. THE LYMPHATICS AND NODES OF THE ARM AND AXILLA. The arm is abducted to a right angle with the body—may be viewed from the long side of the page. *A*. The superficial lymphatics and their relation to the veins and axillary nodes Note the assemblage of nearly all of the superficial lymphatics at one point in the lower axilla. *B*. The deep lymphatics and their relation to the principal artery. The superficial lymphatics join them in the lower axilla. The course of the lymphatics follows Rouvière's descriptions

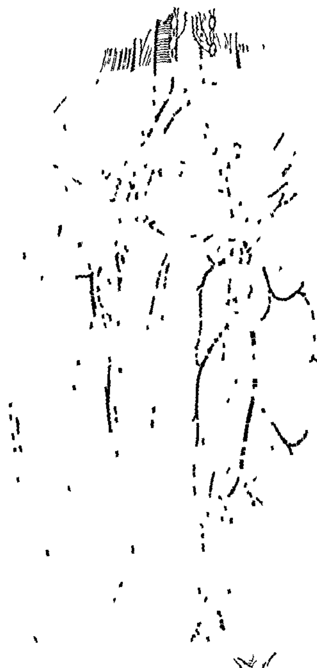


FIGURE 23 THE LYMPHATICS OF THE LOWER LIMBS In the left leg, the superficial lymphatics and nodes are shown, especially their relation to the superficial veins In the right leg, the deep lymphatics are associated with the principal arteries The lymphatics of the pelvic brim are drawn from a dissection by the late Dr John Warren of the Harvard Medical School. (Courtesy of *Annals of Surgery* 100 815, Oct 1934, and the *New England Journal of Medicine*, 204 1025, May 14, 1931)

dant and few other than the serious tropical infestations of various sorts are able to bring on an elephantiasis by an obstruction at the root of a leg or arm.

Much is now known of the anatomy of the lymphatics but as to one important matter no really authoritative statement is forthcoming. They do not appear to drain the muscles.* Lymph vessels running in the intermuscular connective tissue spaces are recognizable but the circulation in muscle itself seems to be carried on by blood vessels and blood capillaries only. That this is actually so is strongly suggested by the absence of any detectable lymphedema in muscle when the superficial tissues of a limb are grossly distended with fluid and crippled by the fibrosis of elephantiasis. For though the limb is then heavy and unwieldy, the muscles remain normal and function naturally. Nor has it ever been shown, in spite of many attempts, that the accumulated fluids of the surface can be introduced among the muscles and so drained from a limb.

Embryological Considerations.—The tree-like organization of the lymphatics in the extremities is most obvious if they are pictured as budding from a single point at the root of each limb, as Sabin has shown. When this development goes wrong, a malformation of the superficial tissue may occur, giving rise to puffy, spongy thickenings or actual cyst formation. Such malformations, or lymphangiomas, may occupy a part or all of a lip or tongue or cheek or hand or foot, may follow the distribution of several dermatomes, or may even cover a whole limb. In many of these lesions, dilated lymph vessels are evident; in others, irregular spaces without any great collections of tissue fluids are found. By contrast, congenital cystic malformations of the lymphatics do occur, especially at the root of the neck or of a limb, huge multilocular cavities difficult to deal with by surgical means.

Physiological Considerations.—The fluid which bathes the cells of the tissues is derived from blood, that is, from the

* Rouvière states (page 4) that "The muscular lymph networks consist of very fine capillaries which are arranged in a variegated manner around the muscle fibers". He quotes Aagard (1913) in support of this statement, but to the writer Aagard's demonstration is not convincing.

phangitis, and (2) a deeper one corresponding to that of the superficial veins, that is, in the subcutaneous fat or actually lying upon the aponeurosis The accompanying illustration shows how groups of these vessels follow, in the leg, the course of the principal branches of the great saphenous vein and, coming together at the saphenous opening, join the trunks which course along the femoral artery and vein This third and last group consists of only two or three rather larger but still fine vessels which drain the foot, the back and outer side of the leg (through the local lymphatics which empty into the popliteal space) and probably the intermuscular connective tissue A very similar condition obtains in the upper extremity Most of the superficial lymphatics enter the mid axilla in order to pass into the axillary glands of this region, while the deep trunks, which are associated with the various great blood vessels, drain the palm and receive in the lower arm the lymphatics which have already passed through the epitrochlear group of glands An occasional lymph vessel passes into the upper axilla in association with the cephalic vein or even jumps the clavicle

In both the upper and lower limbs, each great collecting tree has a very narrow base, at the groin and axilla respectively, after which a group of large lymphatics, about a millimeter in diameter and varying in size and number, winds about the principal blood vessels, the artery in particular, to join the thoracic duct There is, then, a long bottle neck between groin or axilla, as the case may be, and the great collecting duct which is to pour the lymph into the superior vena cava At such a bottle neck the drainage of any one limb can, theoretically, be rather easily interrupted, especially where the lymphatics pass through the great groups of regional lymph nodes As a practical matter, however, the flow is not often cut off at these points, save by operations performed for cancer of the breast, that is, infections, considering the frequency with which they cause an extensive axillary or inguinal adenitis, rarely lead to lymphedema of a limb Apparently alternative routes and emergency passages are abun

It is hard to imagine any injection of foreign material which does not rupture many lymphatics, but in any case motion is evidently a prime factor in the lymphatic absorption of foreign material; for without it the process is exceedingly slow.

Leaving out of consideration the generalized edemas due to an altered chemistry of the blood, most accumulations of tissue fluids in the legs are of a mechanical nature. That is, the lymphatics are absolutely deficient or (on the functional side) more tissue fluid is present than a normal absorbing system can accommodate or, the fluid and absorbing system being normal, the elastic pressure of muscular action is lacking. There will therefore be included in this chapter congenital malformations, elephantiasis, edemas resulting from thrombophlebitis, edemas related to allergy and to infection, and functional edemas related to injury and disuse.

Lymphangiomas, Congenital Malformations of the Lymphatics

As is the case with the hemangiomas and abnormal arteriovenous communications, the actual growths of lymphatic tissue and the various sorts of congenital malformations so merge into one another that a clear distinction between them is impossible. Almost all subcutaneous swellings of lymph vessels are present at birth, and grow with the body, though they may enlarge disproportionately—particularly the cystic ones. If a localized lymphangiomatous or lymphangiectatic swelling, not present at birth, grows in after years, it is likely to be mistaken for a lipoma or plexiform neuroma. Such an event is very rare.

Malformations of the lymphatics are much less common than the corresponding disorders of the blood vessels. They have been divided into three classes, *simple*, *cavernous* and *cystic*, and for the sake of clearness the various sorts will briefly be described here, whether or not they are likely to be found upon the extremities.

Simple Lymphangioma (Lymphangioma Simplex).—This

arterial side of the capillary bed A part of it, consisting principally of water and salts, re enters the blood capillaries, but plasma proteins and all foreign materials are carried off by the lymphatics, whose task it evidently is to remove from the vicinity of the body cells substances whose presence in abnor

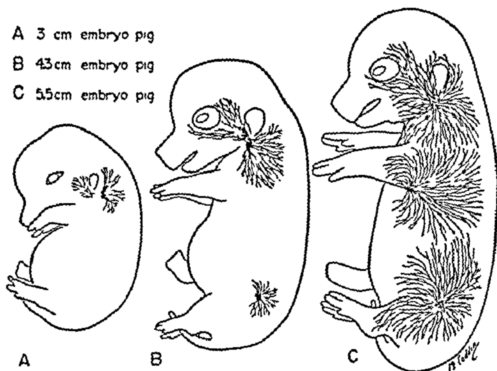


FIGURE 27 THE DEVELOPMENT OF THE LYMPHATICS IN THE PIG Showing how they grow out upon the limbs Modified from sketches of Dr Florence R Sabin

mal amounts would harm the organism Obviously the high percentage of colloidal material in the blood capillaries opposes the entrance into them of extravascular proteins, whereas the lymph capillaries, whose function is purely absorbtive, are quite able to receive them Just how foreign bodies, such as red cells, carbon particles, and other dusts, enter the lymphatics is not understood Of course they can be, and are, carried in by phagocytes, but they are undoubtedly taken up mechanically as well, though whether they are forced through the capillary wall, or infolded by it, or pushed in, after actual rupture of the delicate endothelial cell membrane, is unknown.

oped and afterwards changes little in size, except as it grows with the infant.

Many lymphangiomatous deformities include malformations of the blood vessels, so that it is only possible to say that something has gone wrong with the development of the whole superficial vascular system in the region affected. The surface of an entire limb may be malformed, as a result of which bizarre changes occur. Groups of nipple-like projections from which fluid is discharged at intervals have been described. The writer has encountered a case of this sort in a young girl. At the time of her menstrual periods a brownish fluid would be discharged from these little excrescences, apparently because of some change brought on by the menstrual cycle, a sort of vicarious menstruation. In other cases, the projections appear to consist of local dilatations extruded above the surface, capable of discharging a great deal of fluid, if injured. The boundaries of these cavernous lymphangiomas are vague. They merge at their edges into normal tissues. The back of several fingers may be covered with a soft spongy thickening, which fades out upon the back of the hand. The scar-like, lymph-soaked, vascular tissue of the growth has no clean-cut deep surface but is directly attached to the underlying muscle, tendon, or whatever, without the intervention of the usual aponeurosis. It may even grow into intermuscular planes in the form of wedge-shaped processes.

Treatment of these lesions is very difficult. Excision is the ideal method but anything like a complete, clean-cut removal is rarely possible. Considerable masses can, however, be excised in a series of operations, by turning back flaps of skin, excising the exposed lymphangiomatous tissue down to normal muscle, bone, or whatever lies beneath, and replacing the skin-flaps upon these sound parts. In such operations the blood supply is rather difficult to control and the patient is exposed to the risk of infection. However, by not attempting too much at any one time, much may be done. As an alternative, the tissues may be punctured over many sittings with the endothermy needle and caused to shrink considerably.

sort is confined to the skin and subcutaneous tissues It takes the form of a soft, ill-defined swelling, easily compressible, over which the skin is unchanged Occasionally, very superficial, large, clear lymph vessels can be seen through a very thin cutaneous covering Simple lymphangiomas occasionally appear upon the hands and feet

The writer's experience includes none of these simple lymphangiomas of the extremities but instances are reported of bands of lymphangiomatous tissue passing out upon a limb and containing good sized vessels These vessels, if injured, are capable of causing a long continued leak of lymph, or lymphorrhea

The treatment of such angiomas is surgical excision, or, in case this is impracticable, the use of the endothermy needle Possibly some of them are more or less radiosensitive

Cavernous Lymphangioma (Lymphangioma Cavernosum) —This lesion, which merges into the simple sort, a malformation or growth according to your fancy, is a spongy mass of dilated lymph spaces lined with endothelium It appears as a smooth swelling which may cause great enlargement of such a part as the tongue (macroglossia) or the lips (macrocheilia) On a surface like that of the fingers or the back of the hand it forms a puffy thickening upon which the natural folds of the part may be lost The skin over it is usually normal in color, and the connection with entering vessels is free enough to permit the surface of the mass to be indented However, after many years, especially upon the lower limbs, the tissues may become so fibrosed that pitting is only produced with difficulty With these swellings bizarre deformities of the fingers and toes are often associated, as is shown in one of the accompanying Plates The digits may be of giant size or stumpy or webbed or missing However, the deformed toes or fingers are not necessarily themselves the seat of lymphangioma They may merely exhibit fibrosis and edema A distinction between lymphangioma and a congenital elephantiatic lymphedema is not always clear, but as a rule the elephantiatic part progressively swells, whereas the lymphangioma appears fully devel-



LYMPHANGIOMAS IN CHILDREN. A R F.A, 132543. There is a diffuse swelling of the base of the ring finger and ulnar side of the right hand. A partial excision has already been performed. The left hand is malformed. (Courtesy of [illegible]

tw [illegible]

tw [illegible]

(Courtesy of Dr Augustus Thorndike, Children's Hospital.)

PLATE XIV

Cystic Lymphangioma (Lymphangioma Cysticum) Cystic Hygroma —This form, actually a benign growth, is composed of huge multilocular lymph spaces and is seen more often in females than males Its favorite seat is the root of the neck, but it may appear nearer the jaws, in the axilla, or even, very

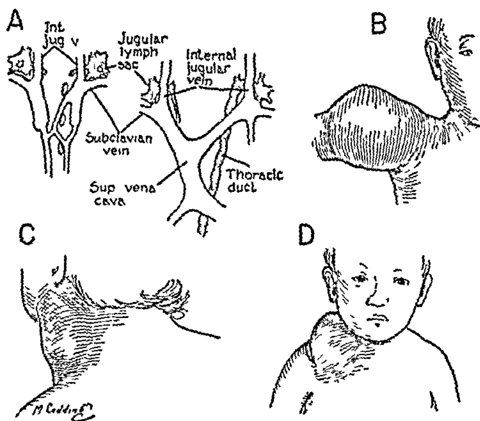
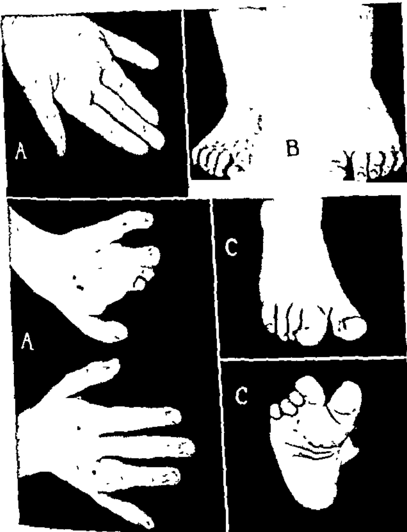


FIGURE 28 HYGROMA *A* The primitive jugular lymph sacs (shaded) of the human embryo and the formation of the thoracic duct (Kampmeier) *B, C,* and *D* The cystic hygroma in various situations Drawn from representative illustrations in the publications of Dowd and of Goetsch

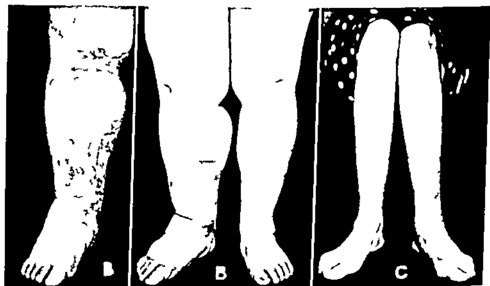
rarely, in the groin It is really a maldevelopment of one of the primitive lymph sacs, especially one of the jugular sacs, which *fails to become united* with the rest of the lymphatic system Its peculiar manner of growth has been explained by Goetsch, who finds, budding from its surface, endothelial sprouts from which microscopic fibrils force their way into adjoining muscle, causing atrophy and fatty degeneration of the fibers they isolate Moreover, the pressure exerted by the



LYMPHANGIOMAS IN CHILDREN. *A*. R.F.A., 132543. There is a diffuse swelling of the base of the ring finger and ulnar side of the right hand. A partial excision has already been performed. The left hand is malformed. (Courtesy of Dr T. H. Lanman, Children's Hospital, Boston.) *B*. J.B.A., 185144. Aged two. A diffuse swelling of the left foot, dorsal surface. *C*. G., A86622. Aged two. A malformation, partly lymphangiomatous, of the right second toe. (Courtesy of Dr. Augustus Thorndike, Children's Hospital.)

PLATE XIV

A Very Extensive Lymph-hemangioma R N M, 45318, a girl, aged fifteen Notice the capillary hemangiomatous patches and the peculiar spots, which discharged a bloody fluid with the catamenia, upon the larger swellings at the groin and on the back of the lower thigh



B I P, 51493 aged 36 Infectious type of Elephantiasis In the central figure, note ulcers on the right On the extreme left this same leg (dark red) during an inflammatory attack Treated by lumbar sympathectomy Since then no more attacks and most of ulcers have healed *C* T H, 20609 (ODD), aged 56 Bilateral edema of an allergic sort Repeated attacks of allergic edema due to fungus

PLATE XV



ELEPHANTIASIS NOSTRA *A* H S L, 50461 A woman, aged thirty-seven From left to right before elevation, after elevation for a week, after second stage, after fourth stage *B* H F W, 56299, a man, aged forty-three From left to right: after elevation for several days, during an attack (dark skin means redness), after several operations. The scars on the thigh are the result of old Kondoleon procedures

PLATE XVI

growing cyst causes it to envelop structures in its path. Yet the hygroma is never malignant.

Being a growth of congenital origin, the hygroma usually appears, more often just above the right clavicle than elsewhere, in the first year of life, and before two years have passed, has become an orange-sized swelling, fluctuant, perhaps translucent, but not to be obliterated by pressure. The accompanying sketches, made after the Figures of Goetsch and of Dowd, will give an idea of the nature of the tumor. Only when it first shows itself, as it occasionally does, in childhood or adolescence rather than infancy, is it likely to be confused with such lesions as branchial cleft cysts, soft or cystic neurofibromas, angiomas, hematomas, or broken down lymph nodes. The fluid contents of the hygroma are thin, clear, watery, and contains as a rule a low percentage of protein, so that it does not coagulate on exposure to the air; and since it is independent of the lymphatic vessels in general, it does not cause swelling of the limb at whose root it happens to be situated. It may, however, force its way into the mediastinum, enveloping the great vessels in its path.

Treatment is excision. Radiation or the injection of sclerosing solutions can not be recommended. Operations, for the most part, are performed in the second year of life. Some are very difficult, especially when the hygroma invades the mediastinum. It is important, if the operation is to leave nothing behind, to excise the entire mass unruptured.

Lymphadenocele.—This name is given to a swelling of lymphatic origin, particularly at the groin, which appears to be derived from cystic enlargement of the regional lymph nodes. Such a swelling may perhaps be related to cystic hygroma or to filarial infections. In the latter case, the female filarial organisms may be found in the lesion. Any cystic disease of the lymph nodes at the root of a limb is of course likely to be associated with a lymphedema of the whole limb, that is, elephantiasis, a subject next to be considered. It is difficult to see how a lymphadenocele can be treated by any method other

than excision and why excision should benefit (or aggravate) obstruction of the lymphatics of the limb involved

Elephantiasis

There are certainly three and probably four varieties of elephantiasis, if one chooses to accept a classification based on the manner in which the disease makes its appearance However, since the means by which lymphatic drainage is put out of action is far from clear even in tropical (filarial) elephantiasis and is utterly unknown in the sporadic and hereditary sorts, classification had better be as simple as possible Accordingly, the varieties presently to be described are distinguished from each other sometimes on pathological and sometimes on clinical grounds without much thought of scientific accuracy

1 **Surgical Elephantiasis (Elephantiasis Chirurgica),** usually results from operations performed for mammary cancer but very occasionally for serious protracted infections or malignant tumors of the lymph nodes themselves Thus, most instances of this sort occur in the arms

Elephantiasis of the arm, following a radical operation for mammary cancer, appears to be related to destruction of the great collection of superficial lymphatics as they enter the axilla to join the deep perivascular lymph vessels It happens in a freakish way, most often perhaps when the regional nodes are already infiltrated with cancer but on occasions as well when cancer is altogether absent, as after operations performed under a mistaken diagnosis Nor does it seem necessary, as Halsted assumed, that infection should be a factor That the lymphatic obstruction is not due primarily to interruption of the perivascular lymph vessels coming up from the lower arm is suggested by the anatomical fact that those lymphatics are related to the artery (and even the great nerves) rather than to the axillary vein Actually, of course, it is the vein rather than the artery which is exposed and cleared of lymphatic tissue in the radical operation upon the breast

It is the writer's experience that elephantiasis of the arm

is most likely to follow a radical mastectomy (extensive cancerous invasion of lymph nodes not being a factor) when a considerable accumulation of lymph has appeared in the operative field during the week or ten days following operation; that is, when there is evidence that a considerable body of lymphatics has been interrupted. Upon cessation of the leak, swelling of the arm is apt to begin. However, swelling may rarely appear, disappear, and recur, as if lymph drainage were teetering on the edge of success for some weeks or even months Once a surgical lymphedema sets in, it is almost always progressive, for when too much of a task is put on a few remaining vessels, there is experimental ground * for believing that they may easily dilate and so become functionless. Moreover, postoperative elephantiasis of the arm shows itself first in the forearm rather than in the hand, whose lymph drainage seems to persist for some time via the deep lymphatics after that of the superficial tissues of the arm and forearm has been interrupted. Recently, Veal has analyzed various types of elephantiasis of the arm and has found that there is a pure lymphatic variety, a sort due to venous obstruction and a mixed type. He regards recurrent cancer as mainly responsible.

A surgical elephantiasis of the arm behaves, in respect to the peculiar febrile attacks so familiar in permanent lymph stasis, about like any other elephantiasis, and is subject, unpredictably, to the onset of these attacks. But because the arm is less dependent than the leg, and more easily drained of its tissue fluid by gravity, its elephantiasis is usually rather less disabling. Only when recurrent cancer aggravates the obstruction by actually blocking veins and tissue spaces as well as lymphatics is surgical treatment likely to be demanded. Then, of course, amputation is almost the only recourse. Should relief be desired in less serious cases, the operation advised for elephantiasis of the leg may be used (page 310). It should be remembered that even though the lymphatics are destroyed,

* The writer has observed an
nearly

fluids still flow by gravity through tissue spaces, but the tissues have now lost the advantage of the valved vessel in forwarding fluid against gravity Fluid, therefore, flows out of the arm on elevation and accumulates in it on dependency Elevation plus exercise drains it most effectively

With surgical elephantiasis should be included the terminal *lymphedemas of the leg associated with malignant growths in* the female pelvis These are clearly due to cancerous invasion of the lymph vessels and nodes along the pelvic brim, as by ovarian or uterine cancer There should also be included the serious edemas which may ensue when a mass of primarily malignant lymph nodes is treated by surgical excision plus irradiation

2 **Elephantiasis due to Infection** —The serious progressive lymphedemas which arise from this cause are apt to be associated with some open lesion such as an ulcer Through this, the infectious orga

interrupt them by

almost always the leg which is affected In some cases, it is difficult to decide which is the cart and which the horse Does the lymphedema arise from repeated infection, or does the elephantiasis occasion, at an unusually early stage, the attacks of cellulitis so characteristic of the advanced disease and so hasten its own development? The leg, in cases of this sort, is apt to be enlarged and tense at all times, having little *tendency to the drainage and wrinkling on elevation which* marks most other elephantiases Moreover, there is a suspicion *of an allergic reaction in some cases, a hypersensitive*ness to fungi or bacteria entering through the lesions of epidermophytosis such as will be described in a later section The elephantiasis based on infection is rare in temperate re

ommon, as Matas has stated,

infection of all sorts, are

approached Under such circumstances, a femoro iliac thrombophlebitis occasionally leads to a disease of this kind Unless open ulcer or evidence of infection clearly forbids plastic operations, such treatment as is used for the commoner sorts

of elephantiasis is suitable for this form. Actually, preventive treatment is the main thing. Fungus and bacterial infection must be fought off by appropriate means. In this respect the writer has more than a suspicion that the vasodilatation secured by lumbar sympathectomy may prove useful.

3. Elephantiasis Tropica, the filarial sort, is due to permeation of the lymphatics of the limbs by the parasite, especially the large female form. Except in those bizarre elephantiases in which a lymphadenocele is present, and except that the arms are sometimes involved and that the secondary febrile attacks are more often observed in tropical countries than elsewhere, there is nothing about this form of the disease essentially different from the hereditary and sporadic varieties. The manner in which the lymphatics are destroyed is different but the end-result is much the same. Considerable numbers of calcified worms can be found in the tissues, as O'Connor, Golden and Auchincloss have demonstrated by the aid of the X ray. These may also be present in individuals who show no sign of elephantiasis. Tissues containing them have been excised by Auchincloss in an effort to remove tender spots and areas which have persistently ached, areas which have seemed to the patients to be starting points for their inflammatory attacks. His removal of such elephantiatic tissues led him to advise operation for this purpose only and not with the idea of securing drainage of the lymphedema as such. Since this general plan of surgical treatment now seems to offer the best chance of success in the nontropical as well as the filarial sorts of elephantiasis, it is presented below.

4. Elephantiasis Nostra: Milroy's and *Meige's Disease.*— These are lymphedemas exactly *similar both* pathologically and clinically, *but for which* no *cause* whatever can be *discovered*. Milroy and Meige described *independently* an hereditary elephantiasis as each saw it in a *family*. Hope and French have written a most picturesque *account* of another family. Almost all of Milroy's cases were *not only* familial but congenital, that is, the *elephantiasis* developed from birth; whereas in the other families the condition first showed itself,

as a rule, in the neighborhood of puberty The disease exhibits all the characteristics of sporadic elephantiasis, its situation, progressive course, tendency to inflammatory attacks and to gross terminal deformities It therefore requires no separate description

Elephantiasis nostra, or sporadic elephantiasis, is not particularly uncommon Certainly it is vastly more often seen than the familial form More would be heard from it were it not that most of its victims are not only ashamed to show their legs but are told that nothing can be done for them Both sexes are affected, females perhaps the more often The onset is usually at about the time of puberty, rarely at birth or during childhood, occasionally after the twentieth year and very rarely indeed after thirty As a rule, the disease is confined to one leg, but perhaps one case in eight or ten is bilateral

As in filarial elephantiasis, enlargement of the leg is gradual The ankle is swollen first, then edema mounts to the knee and finally invades the thigh In the beginning, the skin is unchanged and the superficial parts pit on pressure, but as time goes on, the skin becomes thick and the subcutaneous tissue hard After some years, pitting no longer takes place unless the parts are made to soften and wrinkle by elevation of the leg for several days By the yielding of the skin here and there sacculations of strange shape are made These are apt to be separated by deep creases All changes are less well developed in the thigh then the lower leg and the swelling stops cleanly at the inguinal ligament and the crease below the buttock If a shoe is constantly worn, the foot does not enlarge and the appearance of the skin which it covers does not change That is to say, continuous pressure, in the form of a boot or anklet, keeps the tissues drained of fluid The blood supply of the elephantiatic leg, so far as can be judged by the color of the skin and the surface temperature, is entirely normal, and so it certainly appears at the operating table

The explanation of these clinical findings is to be found in the state of the tissues and tissue fluids both in the experi

mental animal,* in which the disease has almost exactly been reproduced, and in the human patient. As the tissue fluids accumulate, their content of protein increases until it may reach a height of even four per cent—half that of blood serum. With this change, fibrosis advances, perhaps because the highly proteinized fluid acts as a tissue-culture medium. Characteristic coarse trabeculations become visible in soft tissue roentgenograms and constitute, as Reichert has shown, a feature of the disease. All such reactions seem to be proportional to the degree of stasis. It has already been explained that they are almost unnoticeable in the area of foot covered by the shoe and that they are far less marked in the thigh, from which fluid can rather easily escape into the abdominal wall or retroperitoneal tissue, than elsewhere.

Efforts to demonstrate a valved lymphatic, or indeed anything like a normal lymph vessel, in the elephantiatic leg have been fruitless. There may be great spaces containing fluid either just beneath the skin or upon the thickened aponeurosis. But the injection of trypan blue, which the lymphatics readily take up and retain, fails to demonstrate any paths for tissue drainage except such spaces. Sometimes the dye injected into the deep skin will run out rapidly, making a diffuse splotchy stain. Indeed, if the leg is elevated, it may pass up from the foot to the thigh in a few minutes, just as is the case in the elephantiatic leg of the dog, though in other instances, it merely makes a local blue mark and moves hardly at all. If, however, a patient elevates the leg at an angle of thirty degrees for a few days, the fluid may reach the body so rapidly as to form a swelling in the corresponding flank and skyrocket the urinary output. As a rule, if the leg has not been elevated, puncture with a needle anywhere in the calf will obtain a flow of clear lymph, but sometimes the engorged spaces are not reached by the needle, so that hardly a drop of fluid will flow.

The deep as well as the superficial lymphatics are totally absent. The writer has explored the femoral vessels of the

* Drinker, Field and Homans. Also Homans, Drinker and Field. See Bibliography at the end of the chapter.

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strual periods or in connection with a respiratory infection; especially, perhaps, when the patient is tired or run down. In a recent case treated by the writer, the young man, following the usual plastic operation, remained free from attacks (which had previously made an invalid of him) for some two years. Then, during a very damp hot summer, an old epidermophytosis of the foot recurred and he suffered a febrile attack. Probably streptococci entered by way of the fungus infection. It has recently been shown that in the experimental elephantiasis of the dog, the attacks, like those of man, may set in spontaneously. This has offered the opportunity, which Drinker and Field seized, to secure tissue fluid just as the attack reached its height. They found in the fluid, at this moment, with perfect regularity, a limited number of streptococci. The same strain would persist in the attacks for many months, and then mysteriously a new strain would appear. The local heat, swelling, fever, and prostration exactly imitated the human disease. However, soon after the attack had reached its height and between attacks, no bacteria could be found in the tissues. Doubtless in man the attacks are of similar character, and in parts of the world where the disease is common have been treated by polyvalent streptococcal sera with considerable success. Possibly sulfanilamide will prove effective against them and should undoubtedly be tried on the ground that the streptococci are probably hemolytic.

Advanced elephantiasis, associated with extraordinary malformations, frequent febrile attacks and perhaps local ulcerations, is a very serious disease. A case in point is illustrated herewith, that of a man whose attacks had worn him down so that not only did his unwieldy leg keep him from work and all enjoyment of life but he had become dull and depressed, a chronic invalid. Removal of much of the diseased tissue has made a new man of him. Even if his attacks should occasionally recur he will have taken on a new lease of life. It has been shown furthermore that in such a case as his, bacteria are present in the tissues at all times and that operations must be

elephantiatic leg without finding any lymphatics about them. However, on the occasion of a pelvic exploration, in the case of a young woman suffering from elephantiasis of both legs and the external genitalia, the great perivascular iliac lymph vessels were actually observed They were large, by reason of being thick walled, as if chronically inflamed, and actually contained the least bit of lymph, so that some remnant of lymph flow remained However, it could not be determined what tissues they were draining (probably those of the pelvis only) and the lymph nodes were curiously flattened and atrophied Whether such an appearance is the rule is unknown

The Febrile Attacks of elephantiasis are dramatic and peculiar They rarely appear until the disease is well advanced Many persons are free from them altogether Indeed, in both the sporadic and hereditary varieties of the disease, which are alike also in other respects, perhaps not more than one patient in four suffers from this complication However, once attacks are established, they continue at long intervals or short as the case may be In a typical instance, the elephantiatic leg first feels uneasy and within a few hours becomes hot, red, and additionally swollen The temperature rises so rapidly that a chill, often a very severe one, is almost always felt The infection is distinguished from others by involving the whole limb at one time, not in a creeping manner The temperature rises to 102°–104° F (39°–40° C) and the patient is often severely prostrated But for all that, the attack is self limited It usually lasts for three to five days, being totally uninfluenced by treatment

The cause of the attack, which is far more common in the filarial elephantiasis of tropical countries than in the elephantiasis nostra of temperate climes, has always seemed obscure It is usually dubbed erysipelas or cellulitis, yet bacteria have seldom been recovered from the tissues Moreover, the timing of the attacks has always been erratic They may occur about once a month or once in six months, with men

lar spaces beneath the aponeurosis. To that end, long incisions were made, flaps were turned back, lymph-soaked tissue and aponeurosis were removed and the skin flaps were replaced upon the muscles. It was thought also that pathways could be made to lead fluid to the body by prolonging the incisions from the leg over the hip to the flank or over the inguinal ligament in front. These latter schemes were based on Handley's plan of inlaid silk strands which were intended to drain the elephantiatic arm after radical mastectomy. Unfortunately there is no such thing as restoring lymph drainage, nor does a scar conduct fluid to the body as effectively as nature's widened subcutaneous tissue spaces. Thus any benefit which ensues upon plastic operations seems to be due to the removal of those tissues in which the fluid is formed and retained. The ideal procedure is, then, the making of the thinnest possible flaps of skin, removal of all soft parts down to healthy, muscle, tendon and bone—none of which seem to accumulate tissue fluid—and replacement of the skin-flaps upon these deep structures. Sistrunk went a good way toward accomplishing this, Auchincloss frankly planned to do away with (filarial) elephantiatic tissue and the operation described below seems to go about as far in the way of reducing the amount of subcutaneous tissue as it is possible to go. Moreover, since the lower leg is the chief reservoir of fluid and is subject to the greatest deformity, there is no reason in most cases for carrying the plastic higher than the knee. Actually the thigh is reduced in size by the complete operation below, for less fluid now has to pass through it on the way to the body, and the thigh itself is rather readily drained by elevation.

The accompanying sketches will clarify the following brief description. Under a general anesthetic, the leg, foot, and toes are thoroughly cleansed by any routine method. Then, when everything is ready to procede, an Esmarch bandage is applied in a broad band to the thigh. This may have to be reinforced by rubber tubing, for the elephantiatic thigh is compressed with difficulty and the operative field must be bloodless.

An incision is then made from just below the knee to the

performed with great care No sort of incision, much less an elaborate plastic, can be made in his case without exciting infection and sloughing (Plate XVI B)

Non-Operative Treatment—On the ground that fluid can escape from the elephantiatic leg only by way of tissue spaces, it would seem that unless the individual spends his life with his feet higher than his head, his leg must gradually enlarge Such is not quite true or at least not true in all cases The wearing of a firm anklet and bandage will keep some legs fairly free from accumulated fluid and the shoe is so effective that the same size can usually be worn on the foot of the elephantiatic leg as on the other If the individual, in addition to receiving the aid of this sort of pressure, is able, when sitting, to elevate the leg upon a stool or chair, for a good part of the day, and does not have to stand for long hours, drainage of the leg is still further favored Finally, the foot of the bed should be raised, if possible, six inches, so that gravity may help to empty the limb at night The writer knows of a working girl who, having invented these means for herself, has used them so successfully as to be able to control the swelling of her leg Possibly her tissue spaces are particularly adapted to such treatment, but she can actually pass an evening in dancing provided she is willing to use a little extra elevation before and after Of course, a leak of lymph, such as may follow any little injury, keeps swelling down, but the excessive wetness is not exactly compatible with normal activities

Fungus infections must be overcome They offer an entrance to bacteria and in that way, as already explained, may occasion the streptococcal attacks Ulcerations, if present, are a similar menace However, in any but the very advanced case they are not apt to occur, for, in temperate regions at least, most elephantiatic tissues heal about like any others

Operative Treatment—There has been a good deal of misunderstanding about the objects of the sort of plastic operation, based on Kondoleon's original procedure, which has usually been performed Kondoleon's idea was to drain fluid from the lymph soaked superficial tissues into the intermuscu

in it the row of little perforating arteries and veins given off by the anterior tibial vessels, lateral to the tibia. Then the incision is carried down through the muscular aponeurosis so that the flaps, when turned up, lay bare periosteum, muscle, tendon-sheath and even the capsule of the ankle joint. Superficial nerves are as a rule ignored but the posterior tibial vessels and nerve are carefully avoided.

The great flaps are turned back so as to expose more than half the circumference of the leg. Then the thin skin-flaps are defined. For perhaps two-thirds of the width of each, the flap is formed of bare skin, the deep skin being cleaned just as if a whole-thickness graft were being made. However, at the base of each flap some fat is left, particularly where draining veins are visible, for the difficulty with these flaps is that they have too little venous drainage, not too little arterial supply. The flaps on the foot are made less completely of skin than those of the calf. Just how much of the edge of each flap to remove is a problem. Much extra skin is of course present, and it would seem that the final tension upon each flap must resemble that of the skin in its original state (just as is the case with a full-thickness skin graft).

Once the skin-flaps are made, the fluid-soaked tissues beneath are removed, the base of the great masses being divided with heavy scissors. At this point, the Esmarch bandage is released from the thigh and all bleeding points are picked up with fine snaps and ligated with fine silk, the object being to leave the least amount of injured tissue possible. The field must be left particularly dry if the flaps are to heal ideally. The best results are secured by tacking them down with rows of fine silk stitches, one line in the middle of each flap and the third uniting and holding down their edges, so that very few skin sutures are needed. It will be well to score the flaps lightly near their edges with a sharp knife, which drains their venous blood, just befo [illegible]

the broad trough left by the plastic, held in place with a firm muslin or semi-elastic bandage and supported by a light plas-

foot, having at the top a broad symmetrical Y and at the bottom an inverted Y of which the forward arm is much longer than the other This lower inverted Y is placed below the internal malleolus The incision is made upon the anterointernal face of the leg so that the base of the lateral flap will have

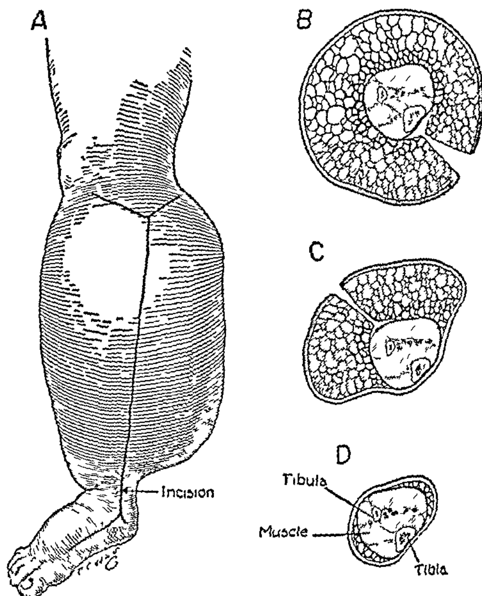


Figure 29 The Two Stage Operation for Elephantiasis *A* The first plastic (antero medial) *B* Cross section of leg showing what is to be removed (shaded tissues) at the first stage *C* End of first stage and plan for second *D* End of second plastic (Postero lateral incision)

be supposed that the lymphatics are not seriously diseased or perhaps that only some particular groups are affected. One can easily conceive that the sort of perivascular inflammation described in Chapter VI as so often engulfing the principal vessels of the limb, may destroy or cripple some of the lymph trunks which wind about the femoral and iliac artery and vein. On the other hand, a moderate venous stasis might keep the tissues soaked in a fluid which only the lymphatics could carry away (high protein content). This might be just too much of a task for them and edema would naturally result. Until more is known of the vasospasms and venous valvular deficiencies which follow thrombophlebitis, an authoritative explanation of this sort of edema can not be offered. However, the occasional appearance, after a femoro-iliac thrombophlebitis, of signs of vascular spasm or even a reflex dystrophy of the extremity, suggests that a vasomotor disorder may be vicious and persistent.

Treatment.—Some regard postphlebitic edemas as purely of venous origin: others, as lymphatic. Actually, for purposes of treatment, the distinction is not very important. The problem is to permit the individual the greatest amount of activity with the least possible resultant swelling: first, because a swollen limb is a nuisance and second, because continued edema encourages fibrosis and the postphlebitic indurations and ulcers already described. Compression by the semi-elastic cotton bandage or the elastic stocking is very valuable, but the individual, like the sufferer from elephantiasis, must plan his or her life for a due proportion of periods of elevation of the limb. And of course any varicose veins which may have developed from collateral venous enlargements during the period of actual obstruction may require surgical treatment. Here the recognition of incompetent communicating veins is very important. Injection is usually contraindicated, and painstaking excisions of the veins is, as a rule, demanded. A description of the diagnostic and technical procedures required for the treatment of such varicose veins will be found in Chapter V. In occasional instances, when evidence of vascular

ter cast This support is necessary both to prevent foot drop and unexpected strains on the flaps

Originally, the writer advocated a four stage procedure, treating one-fourth of the leg's circumference at each stage but the two stage procedure saves much time and seems to result in no serious sloughs Even the necrosis sometimes shown by the skin in patches at the edges of the flaps is apparently shallow However, the operation can doubtless be improved upon by the thoughtful surgeon

The second stage is performed after an interval of perhaps two months, when it may be expected that new flaps can safely be made with their bases toward the old ones The incision now is postero external and since a part of the aponeurosis upon the lateral surface of the leg is inseparable from the underlying muscle, a little of the actual tendinous muscular sheath is necessarily cut away The sural nerve is saved if possible, since it supplies the lateral side of the heel and foot Otherwise the operation is similar to the first one Possibly the whole procedure ought to be divided into three stages instead of two, for then the three sets of perforating arteries, and roughly corresponding veins, could be preserved—the antero lateral (anterior tibial), antero mesial (posterior tibial) and postero lateral (peroneal) A supporting bandage is worn during the first few weeks following the final procedure The patient must of course continue to avoid long hours of standing and elevate the leg whenever possible

Lymphedema following Thrombophlebitis—The local edemas, indurations and ulcerations which follow phlegmasia alba dolens (femoro iliac thrombophlebitis) were described in Chapter V, so that they could be compared with varicose ulcers The sort of edema referred to here is that which, in occasional instances, follows this same form of thrombophlebitis and may rarely persist throughout life It is easily distinguished by the history and by the fact that it affects the entire limb It differs from elephantiatic lymphedema in that it is not progressive but stationary, increasing by day and nearly or entirely disappearing at night It must therefore

after weeks or months in bed, or if muscles or joints are so injured that normal muscular contractions and motions are not permitted, fluid accumulates. Lymph flow, like the flow of venous blood, as already explained, greatly depends on motion and is rather readily disturbed.

Much of this sort of disability can be avoided or at least lessened by exercises and massage during the period of a serious illness and by an arrangement of apparatus during the treatment of sprains and fractures such that the function of muscle and joint can be preserved. Nevertheless the sprained ankle, unless skillfully treated by adhesive strapping and active exercise, may result in many weeks of swelling.

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spasm or reflex dystrophy is present, an interruption of the reflex pathway by periarterial or lumbar sympathectomy is indicated

Lymphedema resulting from Allergy and Infection—It is difficult to segregate the lymphedemas of this sort they must be considered very similar to the swellings of the lips associated with chronic infections of the sinuses, which Stevens described some years ago and with which those dealing with allergic manifestations are doubtless familiar As a result of repeated attacks of infection or allergic swelling, a leg or legs may gradually enlarge, becoming progressively indurated but not ulcerated Such a state, resulting in elephantiasis, has already been described The writer has the impression that the fungus infections may be responsible for some of these swollen legs and the streptococcal infections which enter by way of the lesions of epidermophytosis for others It is very difficult to separate actually infectious from allergic attacks and the matter is all the more confusing in that the protein of dead fungi or of dead bacteria may equally well constitute the exciting cause in any one case

Treatment of such an indurated swelling includes a thorough study of sources of infection and of the possibilities of hypersensitiveness to fungi and bacteria Desensitization may then produce good results Surgical treatment, save for the possible usefulness of a permanent sympathetic vasodilatation in overcoming a chronic cutaneous infection, has little to offer

Lymphedema related to Injury and Disuse—Such must be described as functional and for the most part temporary They *should not be confused with the "traumatic edemas"* related to the dystrophies and causalgias described in Chapter IV These latter are arterial in nature, whereas the sort described here are due to a failure of muscular effort to aid the lymphatics in the disposal of tissue fluids

The borderline between swelling and no swelling is a narrow and delicate one The circulation of the lower extremities has had to adapt itself to the upright position which imposes an unnatural burden upon it If, therefore, the muscles atrophy,

after weeks or months in bed, or if muscles or joints are so injured that normal muscular contractions and motions are not permitted, fluid accumulates. Lymph flow, like the flow of venous blood, as already explained, greatly depends on motion and is rather readily disturbed.

Much of this sort of disability can be avoided or at least lessened by exercises and massage during the period of a serious illness and by an arrangement of apparatus during the treatment of sprains and fractures such that the function of muscle and joint can be preserved. Nevertheless the sprained ankle, unless skillfully treated by adhesive strapping and active exercise, may result in many weeks of swelling.

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CHAPTER IX

INTERPRETATION OF SOME SIMPLE OBSERVATIONS UPON THE CIRCULATORY DISORDERS OF THE LIMBS

In the opening chapter of this book, the various circulatory disorders were named and roughly sorted out. Methods were described by which, once their nature was suspected, they could be studied; how, for instance, an arterial deficiency could be ascribed to arteriosclerosis, to thrombo-angiitis obliterans, or to peripheral arterial spasm, recurrent or chronic. No serious attempt was made, however, to discuss the various possible meanings of common symptoms. Actually, the circulatory diseases of the limbs have a rather limited number of ways of showing themselves. They cause pain or numbness, coldness or heat, cyanosis, pallor, swelling, atrophy, or gangrene, usually a combination of several of these changes. It is by the manner of onset of such symptoms and signs, their combinations and their background of age, sex, and associated disease that a diagnosis is unmistakably indicated. Widely different disorders may sometimes look much alike; arterial embolism and venous thrombosis, for instance. Even those most familiar with the meaning of outspoken, familiar signs may be confused. All the more will those who rarely meet them find certain appearances meaningless and bewildering. It is proposed, therefore, to discuss here some common presenting complexes, showing how, by relatively simple observation and reasoning they may be explained.

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Pain

Pain, as a rule, means arterial deficiency. If it comes on as a result of exercise and disappears on rest, it represents

intermittent limp, and indicates thrombo angiitis obliterans or arteriosclerosis In either case, it may be referred to the calf or shin or foot and will vary in severity from numbness, through cramp, to a sharp stab It is thought to be due to an accumulation *of certain metabolites in the tissues*

Pain which comes out of a clear sky, an agonizing persistent *pain*, usually *means sudden* arterial closure Several causes of such closure should come to mind, depending upon the circumstances under which it appears With all sudden arterial closures, absence of the peripheral pulses, coldness, local paralysis, marbled bluish pallor, and very often some slight degree of swelling are likely to be associated

1 Arterial thrombosis (Chapters II and III) The patient will be one who, because of advanced age, is subject to arteriosclerosis *or whose earlier age, sex (male) and story of intermittent* limp are consistent with thrombo angiitis obliterans

2 Arterial embolism (Chapter IV, last part) The patient, who may be of any age from youth on, will exhibit a fibrillating heart, evidence of mitral stenosis or coronary disease, or of acute cardiac decompensation Immediately premonitory numbness or coldness will often have occurred

3 Venous thrombosis, in the femoral or external iliac vein (Chapter VI) Usually an operation, or injury, or debilitating disease, or childbirth, will be a background for the thrombosis This form, phlegmasia alba dolens, is often ushered in by severe pain, referred to the inner face of the thigh, the *back of the knee or the calf, an indication* of ischemia due to reflex arterial spasm The peripheral pulses may be weak or absent, the leg cold and white Such a state is apt to be temporary—*being followed in a day or so by the characteristic* swelling—and only rarely goes on to the full state of coldness, *mottled pallor*, and final gangrene, with no or little edema, *characteristic of the organic occlusions*

Arterial thrombosis, arterial embolism, and arterial spasm secondary to venous thrombosis need not necessarily cause pain *Coldness, numbness, and paralysis may be the initial*

signs. The gangrene which so often follows, tends to be of the moist type.

Pain of a persistent continuous sort, especially in the toes and forefoot, associated with cyanosis and a shiny swelling, together with some degree of gangrene of one or more toes, indicates a serious arterial deficiency and threatens loss of toes or even foot and leg. The most severe pains are due to thrombo-angiitis obliterans, but arteriosclerotic cyanosis and gangrene sometimes cause great suffering. The pain is usually made worse by elevation and is diminished, temporarily at least, by moderate depression of the foot. The diabetic gangrenes seem to cause little pain (Chapters II and III).

Cyanosis, Pallor, and Coldness

Cyanosis, pallor, and coldness will be discussed together because they are due to a much retarded and usually restricted circulation. Pallor represents constricted vessels; cyanosis, a slow current, irrespective of whether the cause lies in the arteries or veins (Chapter I).

In the fingers (rather than the toes) coming on acutely and temporarily in response to cold or emotional upsets, these signs are due to Raynaud's Phenomenon, that is, spasm of the digital arteries (Chapter IV).

The sudden establishment of cyanosis, pallor, and coldness in any limb, whether or not preceded by severe pain or merely by numbness and paralysis, indicates sudden arterial occlusion, which, as already explained, may be embolic, thrombotic, or spasmodic (associated with thrombophlebitis). If the coldness persists, if the pulses below the root of the limb disappear, if the mottling becomes a fixed purple, and if these changes affect the lower half of the limb, such changes are irreversible and gangrene will follow. Swelling is variable, but the blue, cold area is at least full and often blistered.

Explanation. That some degree of cyanosis and even edema is usually associated with sudden arterial stoppage need not cause surprise. The veins of the terminal parts may even be

prominent When the arterial stream is slowly cut down until it entirely ceases to supply the foot, for instance, the leg gradually atrophies and is, so to speak, drying up even before the final stage of gangrene On the other hand, when blood flow is rapidly cut off, the tissues are caught wet Blood accumulates in the finest blood vessels, having nothing to push it along, and losing its oxygen, while the leg is still warm, gives the skin a deep purple color Very likely the oxygen want due to the sudden arterial stoppage causes the fine vessels to dilate, so that they accumulate the last of the blood delivered to them Actually there seems sometimes to have been an escape of blood into the subcutaneous tissues, as in post mortem lividity, for often the purple color of the mottlings or large patches can not be altered by pressure But even if pressure causes the purple skin to become pale, such a change should not be interpreted as proving that a circulation exists and that the tissues are alive The stagnant blood will flow back into the pale spot with a speed proportional to its accumulation in the neighborhood Only if the limb is elevated, can the manner in which a spot, blanched by pressure, regains its color, be interpreted as meaning anything at all If, while the foot is *elevated,* the blanched spot regains some degree of color, then the circulation is still going on and the part is alive (For a further interpretation of color see Chapter I)

The occasional absence of pain, when a large vessel is suddenly occluded, is very puzzling Any leg which rapidly becomes numb, powerless, more or less edematous, blistered, and pulseless, which takes on within twelve hours a mottled, bluish whiteness, and which, at the end of thirty six hours, appears partly white, partly livid, and utterly cold, has certainly suffered a sudden shutting off of its arterial stream That this can happen without pain is certain, yet the ischemia of an embolic arterial occlusion, when painful, is as agonizing as anything encountered in medicine Morphine may have no appreciable effect upon it

Chronic Cyanosis and Coldness of the Hands and Feet are rather common *In young people,* usually but not always fe-

males, this indicates continuous arteriolar spasm. Hands and feet are equally affected. Excessive sweating of the cyanotic parts is almost the rule. Obviously the state is therefore one of continuous sympathetic irritability, since the parts affected are those most responsive to sympathetic changes. Warm surroundings lessen but never abolish the cyanosis; they merely change the tint toward red or even pink. Exposure to cold and emotional strains intensify the sympathetic irritability. The peripheral arteries present normal pulses. In the presence of such a state, sympathetic paralysis, however produced, turns the skin warm and pink; that is, organic changes in the finer arteries have not occurred, and the condition is always curable by sympathectomy.

Chronic cyanosis and coldness *in elderly people* usually means arteriosclerotic deficiency. Peripheral pulses may still be present but more often are absent.

The Peripheral Arterial Pulsations

The Dorsalis Pedis artery is best palpated upon the forward part of the instep, just lateral to the first metatarsal bone, and two to four fingers' breadth proximal to the (distal) head of that bone. This artery is, as a rule, more easily felt than the posterior tibial, and its pulsations are used as the standard for the state of the peripheral vessels of the leg. It almost never happens that, under normal conditions, both dorsalis pedis arteries are insignificant or absent, but if such is the case, the posterior tibials will be particularly strong.

Absence or enfeeblement of one dorsalis pedis pulse is suggestive evidence of arterial deficiency, and in that case the posterior tibial will, naturally, be equally deficient. Such is true of thrombo-angiitis obliterans and all acute arterial obstructions. Absence of the dorsalis pedis is not, however, necessary to a diagnosis of arteriosclerotic or of diabetic arterial deficiency or gangrene. In fact, arteriosclerotic gangrene of a toe occurs fairly often in the presence of a pulse, though usually a weak one, in the dorsalis pedis. This is even more likely to happen in a diabetic, in whom an arterial de-

prominent When the arterial stream is slowly cut down until it entirely ceases to supply the foot, for instance, the leg gradually atrophies and is, so to speak, drying up even before the final stage of gangrene On the other hand, when blood flow is rapidly cut off, the tissues are caught wet Blood accumulates in the finest blood vessels, having nothing to push it along, and losing its oxygen, while the leg is still warm, gives the skin a deep purple color Very likely the oxygen want due to the sudden arterial stoppage causes the fine vessels to dilate, so that they accumulate the last of the blood delivered to them Actually there seems sometimes to have been an escape of blood into the subcutaneous tissues, as in post-mortem lividity, for often the purple color of the mottlings or large patches can not be altered by pressure But even if pressure causes the purple skin to become pale, such a change should not be interpreted as proving that a circulation exists and that the tissues are alive The stagnant blood will flow back into the pale spot with a speed proportional to its accumulation in the neighborhood Only if the limb is elevated, can the manner in which a spot, blanched by pressure, regains its color, be interpreted as meaning anything at all If, while the foot is *elevated,* the blanched spot regains some degree of color, then the circulation is still going on and the part is alive (For a further interpretation of color see Chapter I)

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Chronic Cyanosis and Coldness of the Hands and Feet are rather common *In young people,* usually but not always fe-

portance are changes from a good pulsation to a poor one or to complete failure. A sudden deterioration is almost invariably a serious sign.

In States of Acute Coldness and Numbness, due to the arterial stupor which occasionally results from direct trauma to a large artery, as by a bullet wound or fracture (especially at the elbow), the peripheral pulses rapidly disappear and may again, though seldom so rapidly, return (Chapter IV). A very special form of spasm of a large artery, the brachial, may very rarely occur suddenly as a result of irritation of its sympathetic nerve supply where it joins the lower end of the brachial plexus in its passage over the highest rib. Usually, nervous symptoms predominate, and the arterial closure is incomplete and chronic.

The sudden disappearance of the peripheral pulses because of arterial thrombosis or embolism, or as a *reflex* with the onset of a femoro-iliac thrombophlebitis has already been described. The ischemia of all such states is usually painful but rarely is a cause merely of coldness and numbness.

Swelling of a Limb, without a Change of Color

Leaving out of consideration cardiac edemas and those due to an altered chemistry of the blood, edema of a limb occurs because more fluid collects in the subcutaneous tissues than the drainage system can carry away. Such an edema may be caused by venous stasis, by muscular atrophy and weakness after an illness, or by loss of the function of lymphatic drainage.

When a whole leg swells from toes to groin within a period of a day or two, especially after an operation or injury, or childbirth, or in the course of a serious illness, but rarely during active life, a femoro-iliac thrombosis is almost necessarily present (Chapter VI). That is, any edema which rapidly mounts to a point above the knee implies that the upper femoral and probably external iliac vein (at least) are thrombosed and obstructed. There may be tenderness over the femoral vessels. The great saphenous vein, secondarily throm-

ficiency is aggravated by a lack of resistance to infection (Chapter II)

The explanation of this distinction between the gangrene of arteriosclerosis and thrombo angiitis obliterans lies in the patchy, erratic character of the arterial narrowing and obliteration in arteriosclerosis Nor are the vessels which furnish the collateral circulation evenly distributed and of regular caliber By contrast, the collateral circulation of thrombo angiitis obliterans, if adequate, is regular and composed of very many fine vessels Thus a patient subject to this latter disease and lacking all peripheral pulses may, unless an especially rapid closure of a good-sized artery occurs, go about for years suffering from an intermittent limp, yet with no obvious change in the nutrition of his toes Whereas an arteriosclerotic sometimes loses a toe or two by gangrene even though the beat in his dorsalis pedis is distinctly perceptible Diabetics, as already explained, are apt to suffer from infection plus gangrene, infection plus necrosis of a phalanx, or infection plus destruction of a toe-joint in the presence of surprisingly good peripheral pulsations

Absence or Enfeeblement of the peripheral pulses in persons over fifty is not particularly unusual Many of these are conscious that they can not walk fast without bringing on an intermittent limp, yet they adapt themselves to this state of things and say nothing about it Their legs are rather atrophied but the color of their toes is unchanged and their toe nails are not deformed On the other hand, some individuals bitterly resent being obliged to walk slowly and persistently seek aid No treatment is likely to be of any great benefit to these, though abstinence from tobacco and the use of Allen Buerger exercises often help considerably On the other hand, they may live to a considerable age without ever losing even a toe

It will be realized that the significance of the peripheral pulsations in the foot is not always easy to perceive The pulses have one meaning in arteriosclerosis, and especially in diabetes, another in thrombo angiitis obliterans Of most im

the area, and a Hufnagel artificial aortic valve was inserted in the first portion of the descending aorta. Approximately four years after the surgery the patient was progressing satisfactorily. He had returned to an essentially normal life, although his cardiac enlargement was noted to have been progressively, but slowly, increasing.

Aortic insufficiency on the basis of a ruptured cusp does not differ essentially from insufficiency from other causes, except for the more rapidly fatal course. The patient frequently experiences a sudden excruciating pain as if something had snapped or torn loose inside his chest. Other patients have immediate fatal syncope, or are unconscious for hours to days. Some patients are immediately aware of a "roaring," "buzzing" or "humming" sensation in the chest. It may be audible to the patient or even to others around him. There may be a latent period, which in the case cited was six days, before the actual rupture occurs following trauma. The diastolic murmur or aortic valve rupture frequently has a musical quality, as was the case in this patient. It has been described as "cooing-dove," "sea-gull" or "humming" sound.

Sudden rupture or perforation of an aortic valve leaflet may occur as a result of bacterial endocarditis (Fig. 318). It is of interest that 25 per cent of the patients we have evaluated for severe aortic insufficiency have had a history of subacute bacterial endocarditis. A number of these had this as a recent complication causing progressive cardiac decompensation, which was one of their main reasons for seeking evaluation. The *prognosis of rupture of the aortic valve during* bacterial endocarditis must vary, despite control of the infection. Among other factors, it will depend on the size of the hole or the severity of the tear. The gravity of the problem makes careful consideration of possible surgical treatment most necessary.

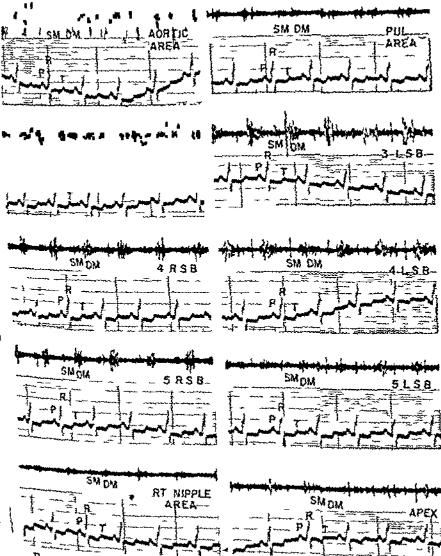
"Right-Sided" Murmurs of Aortic Insufficiency. Although the typical murmur of aortic insufficiency in the great majority of patients is transmitted along the left sternal border to the apex, some [illegible] ransmission along the right sternal border. [illegible] that these patients fall into a group having [illegible]

[illegible]

[illegible] tion of the ascending aorta, and syphilis. Our attention was first called to this small group of patients when we examined a woman who had the typical history and physical findings of severe aortic insufficiency. However, we found that the aortic diastolic murmur was considerably louder along the right sternal border than the left (Fig. 319). Complete evaluation of the patient revealed the stigmata of Marfan's syndrome, and a thoracic aortagram showed aneurysmal dilatation of the sinuses of Valsalva and the root of the ascending aorta. This patient led us to search carefully for "right-sided" murmurs of aortic

sufficiency These soon became evident, confirming the old adage "we find what we look for"

Patients with this "right-sided murmur demonstrated one of the



... to umn, lower tracing) The patient died suddenly

with or without hypertensive heart disease; (2) dissecting aneurysm of the ascending aorta (Figs. 324, 325) caused by Marfan's syndrome or a variant, arteriosclerosis with or without hypertension, or without any evident cause (idiopathic); (3) aneurysm of the sinuses of Valsalva associated with Marfan's syndrome, syphilis or a congenital defect; (4) lesions limited solely to the aortic valve where there are

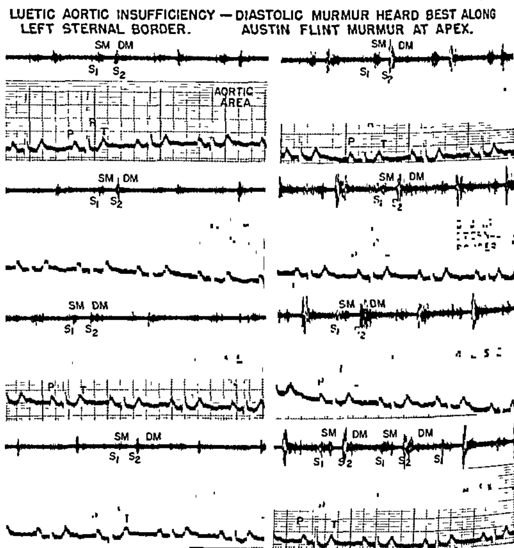


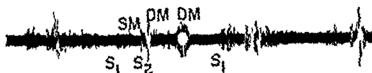
FIG. 322. Woman, age 37. An aortic diastolic murmur (DM) was louder along third and fourth left sternal border (right column) than on right side (left column). Note faint first sound (S_1) and Austin Flint rumble (DM) at apex (right column, lower tracing).

deformed cusps associated with bacterial endocarditis or caused by syphilis.

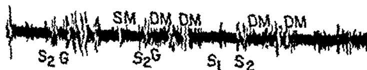
It should be emphasized that the third and fourth right interspaces, and in particular the third (Figs. 319, 320, 321), are the key interspaces. The finding of a diastolic murmur that is loudest at the second right interspace is not unusual in ordinary cases of aortic insufficiency. When the diastolic murmur is loudest along the third right

than of

on either side of the sternum may be ...
except possibly to suggest slight displacement of the aortic root







mitral valve operation was subsequently performed but the patient died nine months later The mitral valve was normal at autopsy

toward the right side. In our experience the presence of an aortic diastolic murmur significantly louder at the third *right* space than the third left interspace has been generally associated with pathologic processes of the types mentioned above. This aspect of cardiac auscultation has been overlooked because it has not been a routine procedure to listen at the right sternal border.

Figures 319 through 321 represent examples of "right-sided" aortic diastolic murmurs that were heard better along the third to fourth right sternal interspaces than along the left. It would appear that in

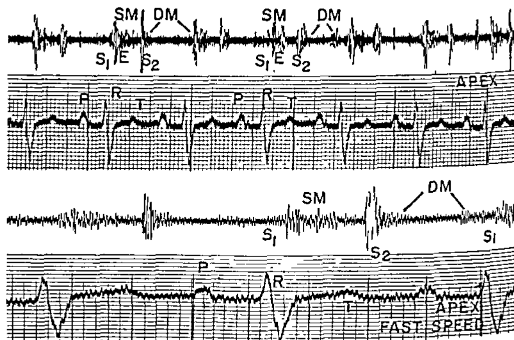


FIG. 324. Man, age 47, with angina pectoris and chronic congestive failure. History of penile chancre 27 years before; positive serology. Cardiac enlargement and calcification of ascending aorta noted on x-ray. Apex: faint first sound (S_1), systolic ejection sound (E), systolic murmur (SM), transmitted diastolic blowing murmur of aortic insufficiency (DM) merging with Austin Flint rumble (DM). Lower tracing shows these events at a faster speed.

these cases there is some process in the ascending aorta or aortic root that causes dilatation and rightward displacement under the right sternal border. It also appears probable that a deformity limited solely to the aortic valve, such as one produced by syphilis or subacute bacterial endocarditis, but unassociated with any apparent displacement of the aortic root, may also transmit the murmur more to the right side of the sternum. On the other hand, any of the above conditions may exist with a murmur that is heard best, as is customary, along the left sternal border. In such instances the murmur of aortic insufficiency is expected to be typically transmitted along the left side (Fig. 322). In addition, although some patients may exhibit some of

the conditions indicated, the murmur will still be better transmitted along the left sternal border. Therefore, it is only when the murmur is heard *more easily* on the right side that one can suspect from auscultation one of these more unusual etiologies.

The Diastolic Apical Rumble Associated with Aortic Insufficiency (Austin Flint Murmur). Although Austin Flint originally described

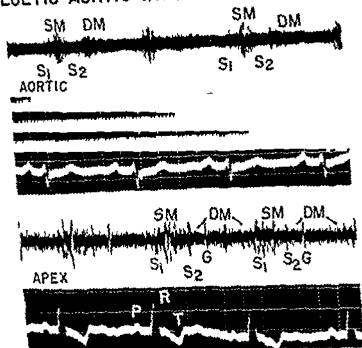


Fig. 325. A 58 year old man with advanced refractory heart failure from syphilitic aortic insufficiency.

a centuation. Postmortem examination consistent with syphilitic aortic insufficiency. Aneurysmal dilatation of the ascending aorta was present. Mitral valve was normal.

the murmur bearing his name as a presystolic m rm n in a n t ant

cumatic fever or other evidence of associated rheumatic heart disease can be clearly designated as an *Austin Flint* murmur (Figs. 323, 324, 325). However, in patients with a rheumatic background (Fig. 326) the differentiation between the Austin Flint murmur and the

toward the right side. In our experience the presence of an aortic diastolic murmur significantly louder at the third *right* space than the third left interspace has been generally associated with pathologic processes of the types mentioned above. This aspect of cardiac auscultation has been overlooked because it has not been a routine procedure to listen at the right sternal border.

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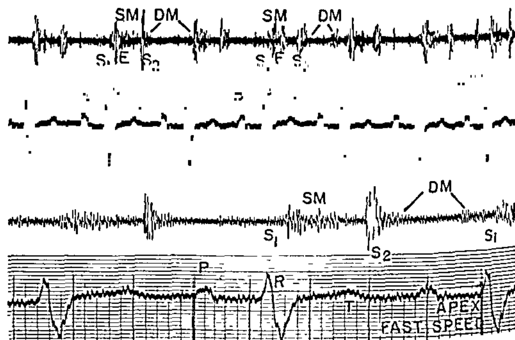


FIG. 324. Man, age 47, with angina pectoris and chronic congestive failure. History of penile chancre 27 years before; positive serology. Cardiac enlargement and calcification of ascending aorta noted on x-ray. Apex: faint first sound (S1), [illegible] M), transmitted diastolic blowing [illegible] Austin Flint rumble (DM). Lower [illegible]

these cases there is some process in the ascending aorta or aortic root that causes dilatation and rightward displacement under the right sternal border. It also appears probable that a deformity limited solely to the aortic valve, such as one produced by syphilis or subacute bacterial endocarditis, but unassociated with any apparent displacement of the aortic root, may also transmit the murmur more to the right side of the sternum. On the other hand, any of the above conditions may exist with a murmur that is heard best, as is customary, along the left sternal border. In such instances the murmur of aortic insufficiency is expected to be typically transmitted along the left side (Fig. 322). In addition, although some patients may exhibit some of

aortic insufficiency including the collapsing type of peripheral pulse, wide pulse pressure, low diastolic blood pressure, and a rocking motion of the heart (seen fluoroscopically) that is associated with increased pulsations of the ascending transverse and descending portions of the thoracic aorta The rhythm is characteristically regular, whereas with mitral stenosis atrial fibrillation is common The first

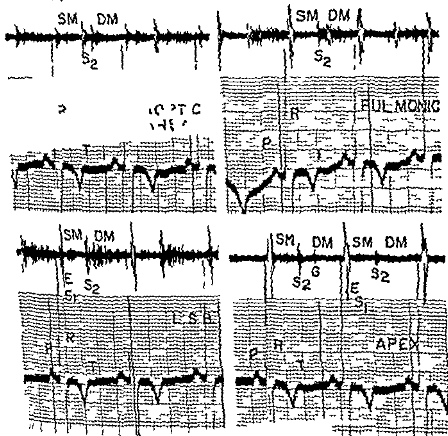


FIG. 323 ...ufficiency Blood pres... ...al valve was explored ... difficult to distinguish between ... sound. Note short aortic systolic murmur (SM) and aortic ... murmur (DM) best heard L S B at apex a ventricular gallop (G) and a ...de III diastolic rumble (DM) were also heard

...heart sound at the apex is generally not accentuated as in mitral stenosis In fact it may be faint (Figs 323 through 326) This is not unusual, as the P-R interval in patients with severe aortic insufficiency is on the longer side and approximately one third have first degree block With mitral stenosis the first heart sound is accentuated. An ejection sound is commonly heard in the first part of systole with aortic insufficiency (Figs 324 326) and is often misinterpreted as

murmur of mitral stenosis has been a more difficult problem. In fact, it has been often stated that one could not make this differentiation clinically, and most textbooks today state that a diastolic murmur heard at the apex with rheumatic aortic insufficiency should be diagnosed as mitral stenosis. However, on the basis of the evaluation of

RHEUM. AI. c̄ AUSTIN FLINT
MURMUR – 2 PTS. (AUTOPSY)

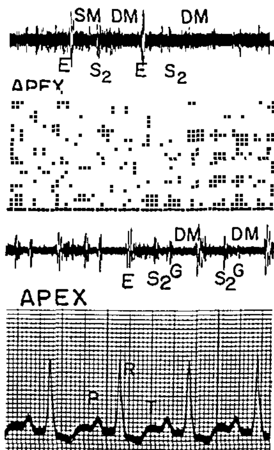
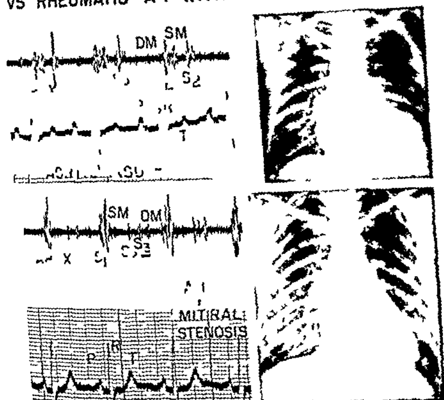


FIG. 326. Two patients with advanced congestive heart failure due to severe

Lower tracing: 32 year old man Two previous episodes of subacute bacterial endocarditis. Blood pressure 160/40-0. At apex had early systolic ejection sound (E), grade III systolic murmur, ventricular gallop (G), transmitted aortic diastolic murmur merging with grade III Austin Flint type of rumble (DM). Died two months after Hufnagel valve operation.

large numbers of patients with severe aortic insufficiency (mainly of rheumatic etiology) it has become evident that one can generally differentiate the Austin Flint murmur from that of mitral stenosis. A total clinical cardiovascular evaluation is necessary to accomplish this. The patient, usually male, with severe aortic insufficiency and an Austin Flint murmur has in addition the classical peripheral signs of

COMPARISON OF FINDINGS AT APEX OF AORTIC INSUFFICIENCY HAVING AUSTIN FLINT RUMBLE VS RHEUMATIC A I WITH MITRAL STENOSIS



bord.r
3ou . . . 92 . old man with rheumatic aortic insufficiency and mitral

mitral valve seen on fluoroscopy The patient d ed postoperatively of cerebral embolus.

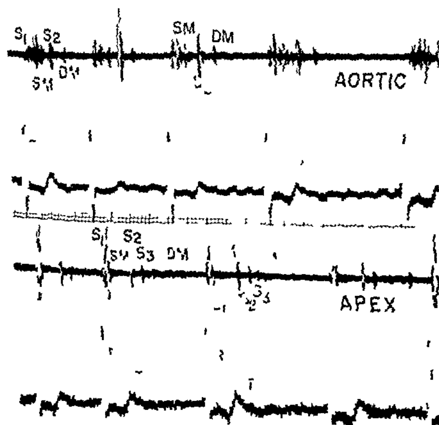
A and notching of the P waves is evident in patient B Straightening of the left cardiac silhouette can be noted in all three patients Figure 330 illustrates the auscultatory findings in another patient with both mitral stenosis and aortic insufficiency

Figure 331 shows the effect of atrial contraction on the intensity and duration of the Austin Flint murmur This 49 year ald man who demonstrated the cla sical findings of aortic insufficiency had a grade

the first heart sound. Sometimes, however, the ejection sound comes so early that it may be difficult or impossible to distinguish it from the first sound (Fig. 327). With mitral stenosis the second heart sound over the pulmonary area is accentuated, and in addition the opening snap is commonly heard in this area as well as at the apex. This gives triple components heard on auscultation, and with inspiration the splitting of the second sound may widen in normal fashion, thereby making these components more evident. These three components would not be heard in an ordinary case of aortic insufficiency. With the typical Austin Flint murmur no opening snap is present; instead, a third sound is heard that comes later than the opening snap. This is a ventricular diastolic gallop (Figs. 323, 325, 326). As a rule, a third sound which has the timing of a ventricular gallop (discussed under Gallop Rhythm) is a point against a diagnosis of mitral stenosis, particularly the "tight" variety. The patient with aortic insufficiency who has an Austin Flint murmur also has a systolic murmur at the apex (Figs. 323 through 326), presumably owing to a relative mitral insufficiency associated with left ventricular enlargement. Three murmurs are therefore heard at the apex: a systolic murmur, a transmitted murmur of aortic insufficiency, and the diastolic rumble of Austin Flint (generally heard best in a localized spot at the point of maximum apical impulse). The diastolic rumble is heard best with the bell of the stethoscope, touching lightly, barely making an air seal, in the same fashion as one auscults for mitral stenosis. The murmur of Austin Flint, as with mitral stenosis, often depends on factors such as atrial contraction, ventricular rate, and rapidity of diastolic filling. The electrocardiogram in patients with aortic insufficiency shows left ventricular hypertrophy, whereas with uncomplicated mitral stenosis there is no evidence of left ventricular enlargement but possible evidence of right hypertrophy. Notching of P-waves is unusual with uncomplicated aortic insufficiency and common with mitral stenosis.

The foregoing points are generally sufficient to make the differentiation between the patient with severe aortic insufficiency who has an Austin Flint murmur and the patient with mitral stenosis. However, individuals who have both mitral stenosis and aortic insufficiency are more difficult to distinguish from those with rheumatic aortic insufficiency and an Austin Flint murmur. As a rule, in the majority, this differentiation usually can be made. Figure 328 shows comparative auscultatory findings at the apex and roentgenograms of the heart in a patient with rheumatic aortic insufficiency and an Austin Flint murmur and in a patient with aortic insufficiency and mitral stenosis. Three patients with combined aortic insufficiency and mitral stenosis are shown in Figure 329. All of these patients had a loud first heart sound at the apex, a third sound that came later than the opening snap of the usual tight mitral stenosis, and a diastolic rumble (which had a presystolic accentuation in patients B and C where a normal sinus rhythm was present). Atrial fibrillation was present in patient

III distinct diastolic rumble at the apex As noted on his electrocardiogram there was a shifting relationship between atrial and ventricular contraction When atrial contraction occurred relatively early in diastole, the atrium being more distended with blood, the Austin Flint murmur had greater intensity and was more prolonged than when atrial contraction occurred later in diastole when the atrium was rela



al on and diastolic rumble"

tively empty and the ventricle more full In some of our patients with

associated mitral stenosis and the ...

Fl

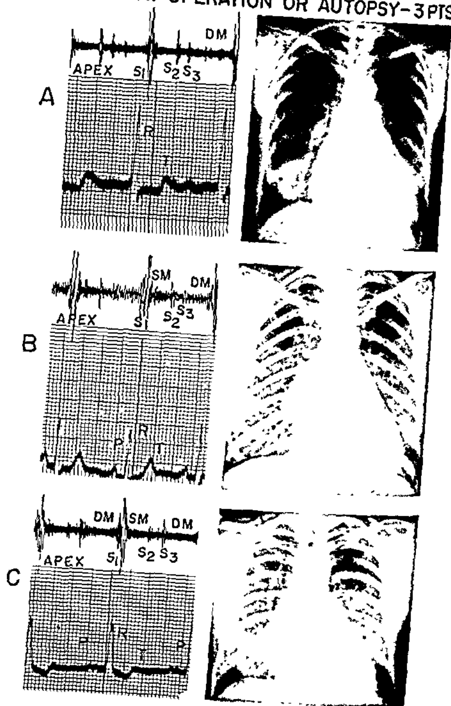


FIG. : rheuma third so blmg m... sinus rhythm). Note notching of P waves in *B*, and straightening of left cardiac silhouette in all patients

AORTIC STENOSIS

Aortic stenosis is generally regarded as the result of a previous rheumatic infection of sclerotic changes (Monckeberg's sclerosis) or of congenital origin With increased knowledge more and more of the cases that previously were thought to be sclerotic are now looked upon as rheumatic or infectious in origin The fact that the aortic valve so frequently became densely calcified misled authorities in their interpretation. There really is no more reason to conclude that a calcified aortic valve originates on an arteriosclerotic basis than to take that view concerning a calcified mitral stenosis or, for that matter a calcified gumma echinococcus cyst or glioma of the brain In many areas of prolonged chronic inflammation, calcium deposits finally develop Furthermore, many of the cases of calcific aortic stenosis, even in elderly patients, have a past history of rheumatic fever or other stigmata of rheumatic infection However, there are instances in which one is forced to accept sclerosis and not inflammation as the primary lesion This occurs especially in bicuspid valves

Auscultatory Findings The diagnosis of aortic stenosis which in previous years was difficult is now being made with a high degree of accuracy With increased knowledge concerning the interpretation of a systolic murmur, and particularly since fluoroscopic visualization of calcified aortic valves has become possible, most cases can now be recognized ante mortem At the bedside the finding of a palpable systolic thrill in the aortic area is extremely helpful It will rarely be due to any other cause The difficulty is that the evidence upon which a reliable diagnosis can be made only becomes clear when the lesion is fairly well advanced A loud systolic murmur does not develop overnight. By the time that a real thrill can be felt, or calcification has developed, the process must have been going on for many years Probably the earliest sign was a slight basal systolic murmur When it is realized that a great many patients have slight basal systolic murmurs without aortic stenosis the diagnosis cannot be made at this stage We have seen quite a few patients who since they had no symptoms or other evidence of heart disease were thought to have an inconsequential and benign systolic murmur—but who reappeared ten to 20 years later with louder systolic murmurs and calcified aortic stenosis. This is further proof that many systolic murmurs are not as benign as generally regarded

The auscultatory findings of aortic stenosis are primarily centered around the interpretation of a basal systolic murmur, though the character of the aortic second sound is of some importance In the absence of such a systolic murmur it will be difficult or impossible to make the diagnosis or even to suspect it Unless the patient is moribund or in advanced heart failure, or has extremely feeble heart contractions or very distant sounds because of emphysema and the like the absence of a systolic murmur practically eliminates the diagnosis

combination of factors plays a part. These factors include the effect of atrial contraction, the time of its contraction during diastolic filling of the ventricle, and the ventricular rate (the rumble being more clearly evident at optimal rates, somewhat analogous to the optimal rate with mitral stenosis). One of the older theories suggests that with significant aortic insufficiency the aortic leaf of the mitral valve is displaced to a position that produces a relative mitral stenosis. Such

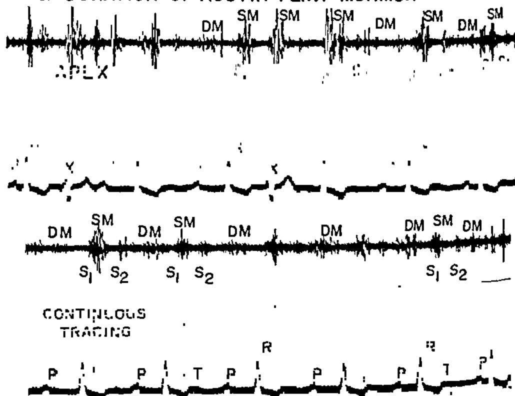


FIG. 331. A 49 year old man with rheumatic aortic insufficiency and Austin Flint rumble. Blood pressure 150/0. A grade IV aortic diastolic blowing murmur was

displacement by the regurgitant aortic stream may play a part, but we have observed patients whose Austin Flint rumbles have remained despite the fact that the diastolic murmur has been reduced from grade IV to grade II as a result of an operation. In addition, we have observed a diastolic rumble in patients with aortic stenosis, particularly those having significant or advanced heart failure. Another possible mechanism is that a relative mitral stenosis results from the excessive enlargement of the left ventricle as compared to the left atrium.

AORTIC STENOSIS

Aortic stenosis is generally regarded as the result of a previous rheumatic infection of sclerotic changes (Monckeberg's sclerosis) or of congenital origin. With increased knowledge, more and more of the cases that previously were thought to be sclerotic are now looked upon as rheumatic or infectious in origin The fact that the aortic valve so frequently became densely calcified misled authorities in their interpretation. There really is no more reason to conclude that a calcified aortic valve originates on an arteriosclerotic basis than to take that view concerning a calcified mitral stenosis or for that matter, a calcified gumma, echinococcus cyst or glioma of the brain In many areas of prolonged chronic inflammation, calcium deposits finally develop Furthermore many of the cases of calcific aortic stenosis, even in elderly patients, have a past history of rheumatic fever or other stigmata of rheumatic infection However there are instances in which one is forced to accept sclerosis and not inflammation as the primary lesion This occurs especially in bicuspid valves

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form. In another case (Fig. 334) a systolic murmur was more prominent at the apex (grade III) than in the aortic area (grade II). Because of this, a diagnosis of mitral insufficiency was made. When the phonocardiogram showed the peculiar diamond form suggesting aortic

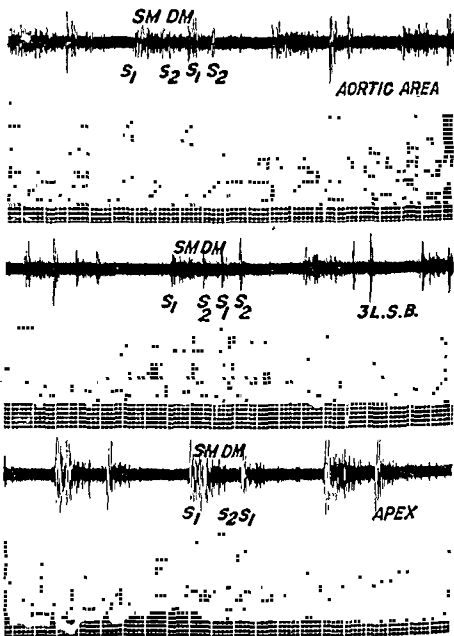
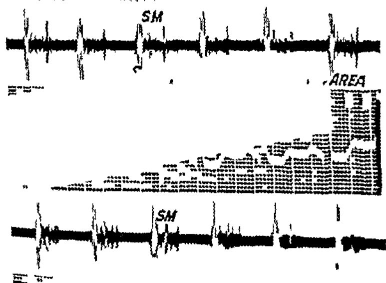


Fig. 338 Woman, age 43, with rheumatic mitral stenosis and insufficiency, and

"diamond-shape" murmur was present, causing some to question the diagnosis of aortic stenosis. At postmortem the mitral valve showed evidence of stenosis and insufficiency, and the aortic valve was normal.

stenosis the patient was reexamined It was then found that, in the upright position during a deep expiration a definite systolic thrill could be felt in the aortic area affording further evidence in favor of the diagnosis of aortic stenosis The systolic murmur is often as loud at the apex as at the base and yet need not indicate that the mitral valve is involved (Fig 335) When the apical murmur has a



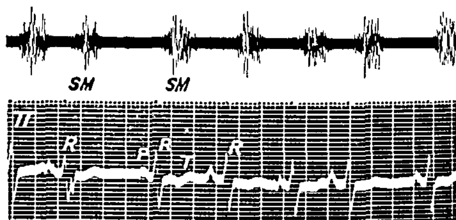
Fi...
nosed b... ...alcific aortic stenosis diag
in the a... ...systolic murmur (SM) was present
throughout... ...a grade V systolic (SM) at the apex (lower
absence c... ...pancy between auscultation and x ray findings and the
patient had ...amond-shape configuration consistent with aortic stenosis the
patient had repeat x ray examination. The calcification proved to be in the mitral
rather than the aortic valve

different configuration as shown in Figure 336 we suspect that mitral insufficiency is also present.

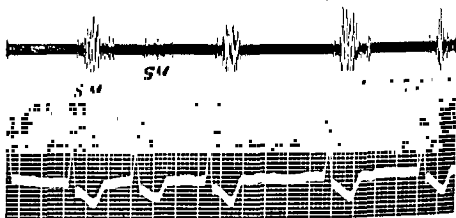
The following experience illustrates that the appearance of the phonocardiogram may occasionally be helpful diagnostically This patient was diagnosed as having aortic stenosis by our roentgenologist because he saw a calcified plaque in the region of the aortic valve (The accuracy of x ray diagnosis when calcification is seen is extremely great as only rare errors have occurred When the phono-

ALTERATION OF SYSTOLIC MURMUR OF AORTIC STENOSIS C̄ "QUICK" BEATS–3 CASES

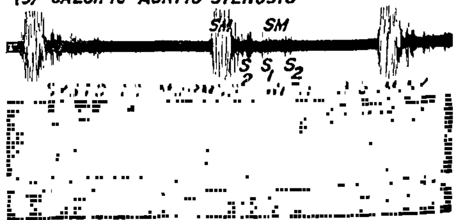
(1) ACCENTUATED FOLLOWING LONGER DIASTOLE



(2) RHEUMATIC AORTIC STENOSIS, AUR. FIBRILLATION



(3) CALCIFIC AORTIC STENOSIS



[pho]nocardiograms were examined, the systolic murmur appeared to have a [crescendo-decrescendo or ...]ntral configuration rather than the characteristic diamond shape [of] aortic stenosis (Fig 338) The clinical diagnosis of mitral stenosis [an]d insufficiency rather than aortic disease was made On postmortem [ex]amination the mitral valve was found to be fishmouthed and cal[ci]fic and the aortic valve was normal. There were also three calcified [pl]aques in the wall of the left atrium the largest one just posterior to [th]e aortic valve. Figure 339 shows another case in which an error in

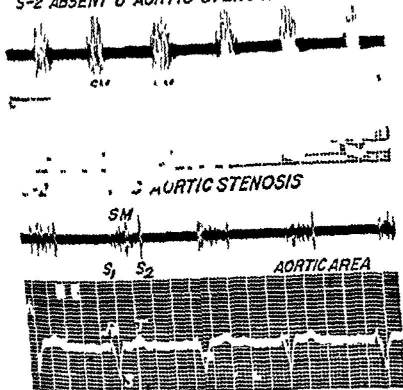


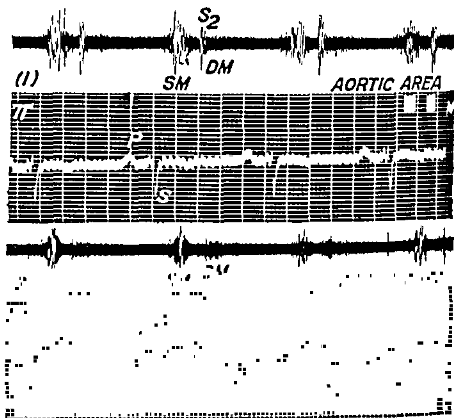
Fig. 341. Upper tracing A 43 year old woman with rheumatic aortic stenosis and insufficiency mitral stenosis and insufficiency Note grade VI systolic murmur (SM) with absence of second sound (S2) Lower tracing Man age 51 with calcific aortic stenosis and atrial fibrillation. Second sound (S2) present.

x ray diagnosis was corrected after the phonocardiograms led one to suspect that the mitral rather than aortic valve was involved

When the rhythm is irregular the systolic murmur in aortic stenosis retains the same characteristics though its intensity may vary It will be fainter with the quick beats and louder after the long pauses whether the irregularity is due to extrasystoles or atrial fibrillation (Fig 340)

It is thought by many physicians that well marked stenosis of a valve must be accompanied by incompetence of that valve In a

previous discussion, evidence was presented to show that pure mitral stenosis without insufficiency was common. The same is true of the aortic valve (Figs. 334, 335, 340). It is a frequent experience to see well authenticated cases of calcific aortic stenosis in which no diastolic murmur can be detected on most careful examination. To be sure, the majority will show the characteristic blowing diastolic murmur of aortic insufficiency (Fig. 342), and when this sound is present,

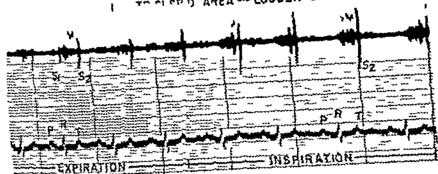


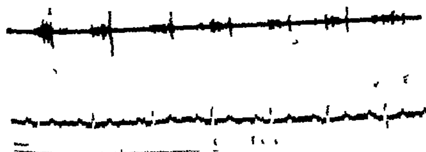
it is an aid in deciding that the systolic murmur is due to aortic stenosis.

TRICUSPID INSUFFICIENCY

Functional or relative tricuspid insufficiency is common. It occurs frequently in all types of heart disease when there is marked right-sided failure. It is particularly common in severe mitral stenosis and in those conditions in which pulmonary arterial pressure is considerably elevated and the right ventricle fails. Under all of these circumstances, a tricuspid valve that is either normal or essentially so be-

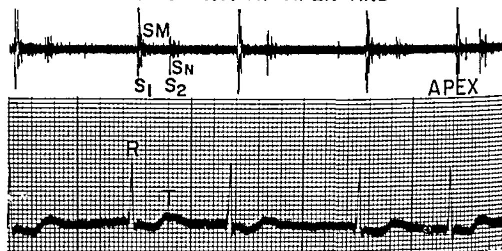
comes dilated because of the increased pressure and size of the right ventricle, and thereby is made incompetent. Organic tricuspid insufficiency ... almost entirely as a rheumatic process and, like mitral ste... ...cy is the presence of a systolic murmur heard best over the lower left sternal region, or sometimes over the xiphoid or at the lower right sternal border. The murmur is pansystolic and is frequently accentuated with inspiration (Carvallo's sign) (Figs 343, 344, 345). This helps to distinguish it





from the systolic murmur of mitral insufficiency with which it might otherwise be easily confused. In contrast, the murmur of mitral insufficiency generally decreases with inspiration, is better heard at the apex, is transmitted well to the left axilla, and if loud enough to the posterior left lung base. The murmur of tricuspid insufficiency varies considerably. It may be soft, high pitched, musical, and sometimes harsh. Although generally heard best along the lower left sternal border, it may be transmitted toward the apex where it can be confused with the murmur of mitral insufficiency. The murmur of tricuspid insufficiency is frequently inconstant, varying in intensity from day to day. As a rule it is louder following exercises. Occasionally it

appears with the onset of atrial fibrillation and disappears with normal sinus rhythm. It is generally loudest following long diastolic pauses, varies with position, and frequently disappears following improvement in cardiac decompensation. The increase in intensity is most marked during early inspiration, becomes stabilized if the breath is held for a moment, and may gradually fall off if inspiration is held.



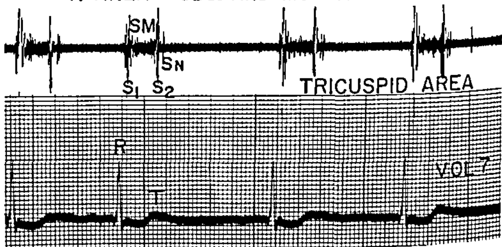
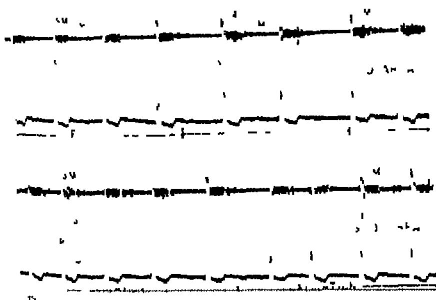


FIG. 344. A 51 year old man with a tight mitral stenosis proved at surgery. No mitral insufficiency. Had pansystolic murmur (SM) heard best over tricuspid area (lower tracing) but also heard at apex (upper tracing).

If inspiration is maintained for a prolonged period, an occasional patient may perform an involuntary Valsalva maneuver which may immediately diminish the intensity of the murmur. This must be guarded against if Carvallo's sign is to be properly elicited. Also, in cases of extreme failure of the right ventricle there may be little or no increase in the intensity of the murmur with inspiration. As already discussed under *mitral stenosis*, a patient with mitral stenosis may

ave a pansystolic murmur heard along the lower left sternal border nd apex that suggests mitral insufficiency Instead, this may well be he murmur of tricuspid insufficiency increasing in intensity with [illegible] (Figs 344, 345)

lition Approximately two years previously, this patient had bee [illegible] an automobile accident where he sustained a crushing injury to his chest. He was now hospitalized for repair of a traumatic aneurysm of the descending aorta Tricuspid insufficiency was present unasso-



with inspiration as shown on continuous tracings.

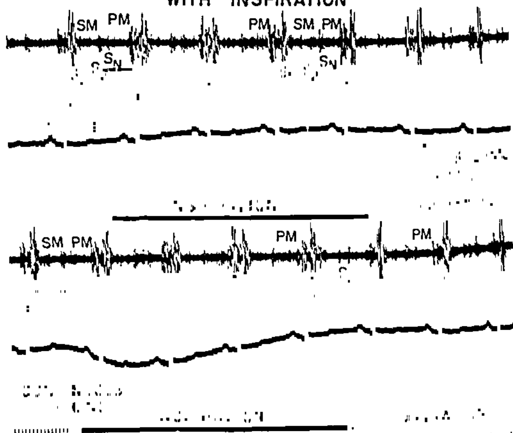
ciated with any mitral stenosis or congenital heart defect This case may well represent a traumatic rupture of the tricuspid valve

TRICUSPID STENOSIS

bu [illegible] of tumor vegetations or thrombus obstructing the tricuspid valve orifice but the great majority are rheumatic in etiology Tricuspid stenosis is often associated with insufficiency of the tricuspid valve, and is less apt to be "tight" than mitral stenosis When tricuspid stenosis is present it is practically always associated with mitral stenosis and therefore in a patient having tricuspid stenosis, differ-

entiation between the murmurs of each individual valve lesion is sometimes difficult. The diastolic murmur of tricuspid stenosis is of the same quality and occurs at the same time during the cardiac cycle as that of mitral stenosis. Its origin is slightly nearer the midline and is usually heard best along the lower left sternal border. It may be well heard along the right sternal border.

When mitral stenosis is also present, the differentiation of the two



tion as shown on continuous tracings at the left sternal border. Faint systolic murmur (SM) and opening snap (SN) also heard.

murmurs by auscultation has been considered almost impossible. However, recent experience with proven cases of tricuspid stenosis where the lesion is a significant one, and particularly if the rhythm is regular, indicates that one can accurately diagnose this lesion. A patient who has auscultatory findings similar to a tight mitral stenosis but without the usual dyspnea on exertion should alert one's suspicion to the possible presence of tricuspid stenosis. A malar flush may be present, and striking prominent "a" waves are noted in the jugular venous pulse in the neck. On auscultation, findings similar to those of

mitral stenosis are heard a loud first sound an opening snap, and a diastolic rumble The diastolic rumble, however, is unusually well heard over the lower left sternal border and over the xiphoid area. Coincident with inspiration, a marked accentuation of the rumble is evident in presystole, doubling or even tripling in intensity (Figs 348, 349) We have recently observed these findings in three successive patients, and have made a clinical diagnosis of tricuspid stenosis

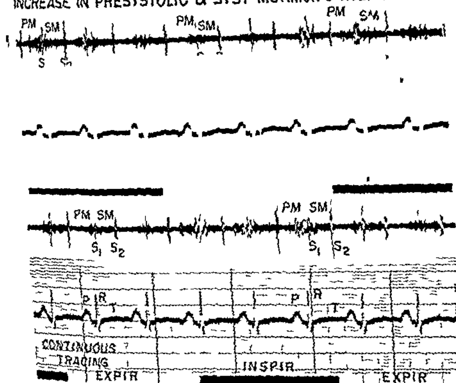


FIG. 348 A 23 year old woman with tricuspid stenosis and insufficiency and mitral stenosis and insufficiency (proved by cardiac catheterization and later at surgery). Presystolic murmur (PM) doubled or tripled in intensity coincident with inspiration, as shown on continuous tracing at left lower sternal border Faint systolic murmur (SM) also noted

Each diagnosis was subsequently verified by cardiac catheterization and/or operation

Figures 346 350 and 351 illustrate two patients admitted for possible surgery for mitral stenosis Both had a history of rheumatic fever but neither had the incapacitating type of exertional dyspnea typical of mitral stenosis Both had a malar flush of the cheeks and prominent 'a' waves of the jugular pulse In both patients, the rhythm was regular and the diastolic rumble was heard well along the lower sternal border and tricuspid area Coincident with inspira

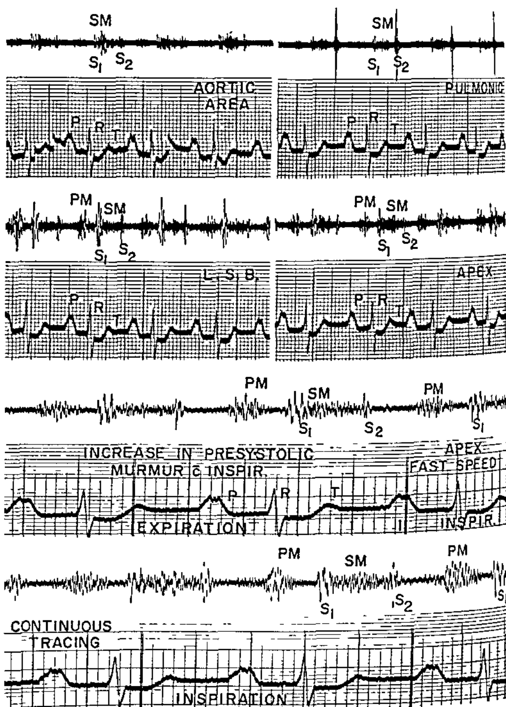
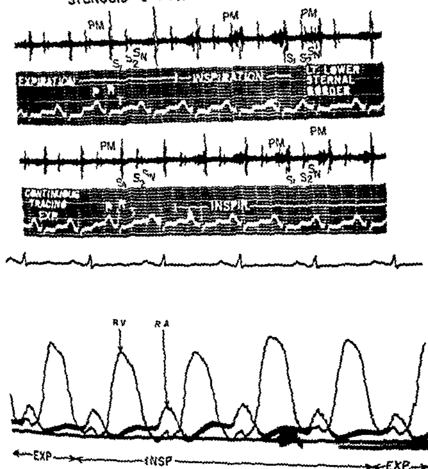


FIG. 348. Woman, age 30, with stenosis of
(proved at postmortem). Note systolic murmur
aortic area (upper left tracing) and also heard well at other areas.
of intensity of presystolic murmur (PM) coincident with inspiration (continuous tracing, lower two strips).

tion the diastolic rumble greatly increased with presystolic accentuation. Tricuspid stenosis was diagnosed and confirmed on cardiac catheterization. Surgery was not performed because of the lack of symptoms at that time



surgery

An opening snap of the tricuspid valve may be present, but in the presence of mitral stenosis it is very difficult to ascertain whether this originates in the tricuspid or mitral valve. Possibly hearing the opening snap better along the lower right sternal border than along the left might be of value here. In general the diagnosis of rheu

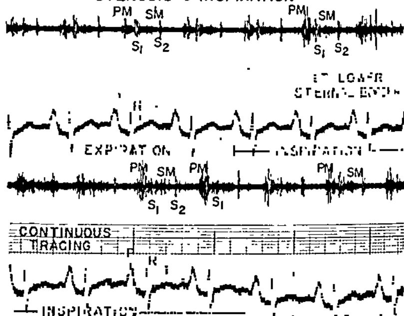
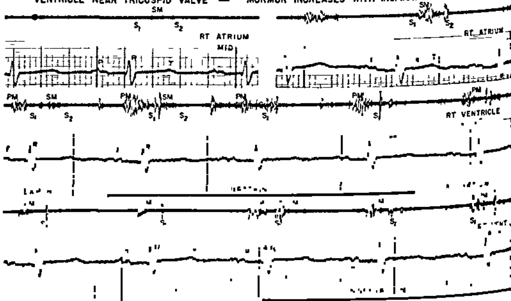


FIG. 350. A 34 year old woman with tricuspid stenosis, aortic stenosis and insufficiency, and mitral stenosis. Had no symptoms of unusual exertional dyspnea. Had malar flush and large "a" waves of the jugular venous pulse coincident with inspiration. Had striking increase in presystolic murmur (PM) coincident with inspiration that was heard along left lower sternal border (continuous tracing). Also had systolic murmur (SM). Cardiac catheterization performed.



... murmur (SM). Presystolic murmur (PM) also heard in a lower area in the right ventricle where it also increased on inspiration (lower tracing). No significant presystolic murmur noted in right atrium (left upper and right upper tracings).

matic tricuspid stenosis is not made solely by auscultation, but rather on the basis of the entire clinical behavior and appearance of the patient. Moreover, many patients with tricuspid stenosis have trival vular disease with stenosis of the mitral and aortic valve as well (Fig 348) Figure 349 shows a patient with tricuspid stenosis proved by catheterization studies and surgery As noted, with inspiration there was increased intensity of the presystolic murmur No significant systolic murmur was present. A third sound heard was thought to be the opening snap With a double lumen catheter simultaneous recordings were taken in the right atrium and ventricle (Fig 349) With inspiration a gradient across the tricuspid valve was noted This corresponded to the increase in the presystolic murmur made across the tricuspid valve, thereby establishing the mechanism of this murmur

It is apparent that an earlier diagnosis of tricuspid stenosis can be made by combining auscultatory findings with other clinical features

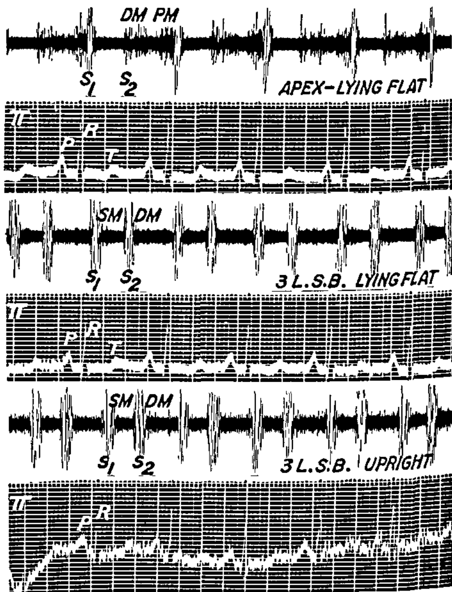
COMBINED VALVULAR DISEASE

(MITRAL, AORTIC AND TRICUSPID)

Valvular lesions are often multiple, and there are various combinations of stenosis and insufficiency of the different valves The diagno is of more than one defect is increasingly difficult but with the total evaluation of the patient utilizing all available methods correct ante mortem diagnosis usually can be made In general it will depend upon criteria that are diagnostic of any one particular lesion bearing in mind the effect the other abnormality may produce As a rule one lesion predominates but occasionally there appears to be an equal involvement in another valve It is obvious that an extremely loud diastolic or systolic murmur in the aortic area may be audible at the mitral area while a faint one would not be so transmitted Additional data apart from auscultation, are often necessary to arrive at the correct diagnosis The presence of dilatation of the left atrium on x ray examination may help to decide that a diastolic murmur at the apex is not transmitted from a louder aortic basal diastolic, but is due to an additional mitral stenosis The configuration of the P wave in the electrocardiograms, the presence or absence of left or right axis deviation or of atrial fibrillation whether the pulse pressure is large or small, may weigh the diagnosis for or against a particular valvular lesion

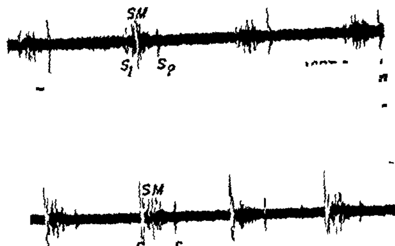
The diagnosis of organic mitral insufficiency in the presence of definite aortic stenosis or insufficiency is particularly difficult It may be ventured if the apical systolic murmur is louder or of different quality at the apex than at the base (Fig 337) and with greater certainty if it is accompanied by definite evidence of mitral stenosis (Fig 352) In many such cases, the apical systolic murmur is really the same aortic murmur and has the same configuration (Fig 353) In fact, the murmur of aortic stenosis is quite frequently heard best at

the apex, and then may be misinterpreted as indicating mitral insufficiency alone or in addition to aortic stenosis. At times, the diagnosis of concomitant minimal or moderate mitral insufficiency in the presence of known aortic stenosis is impossible to make clinically. The diagnosis of mitral stenosis in the presence of aortic valvular involvement will depend on the customary auscultatory evidence, i.e., an accentuated apical first and pulmonary second sound, and a rumbling



urmur in diastole (Fig 354) In the case illustrated by Figure 355, one might have suspected the presence of mitral stenosis in addition to the more obvious aortic stenosis because of the peculiar flat-topped notched P waves and the fact that the first sound was accentuated despite the delayed P–R interval

When the situation is reversed and it is quite clear that mitral involvement is present, the diagnosis of concomitant aortic disease is also a difficult matter If there is a louder and harsher systolic murmur in the aortic area than at the apex, and particularly if there is



hypertension or if a basal systolic thrill is felt, the direction of which is rightward toward the neck or shoulder the diagnosis of aortic stenosis must be entert[illegible] nations of lesions [illegible] nts enabling their

[illegible] notched P waves a third heart sound the timing of which is later than an opening snap, but more consistent with that of ventricular diastolic gallop plus moderate left atrial enlargement leads to the suspicion of associated mitral stenosis in patients with predominant aortic insufficiency (Fig 356) Severe aortic insuf

ficiency was present in the patient illustrated, but because of these associated findings, the mitral valve was also explored at an operation for the insertion of the Hufnagel valve. As is typical in the majority of cases of mitral stenosis associated with severe aortic insufficiency, the stenosis, although present, was not tight, and further opening was not deemed necessary. A grade IV systolic murmur was also present at the apex, signifying concomitant mitral insufficiency. As has been discussed under Aortic Insufficiency, a loud systolic murmur over the aortic area, even in the presence of a palpable thrill,

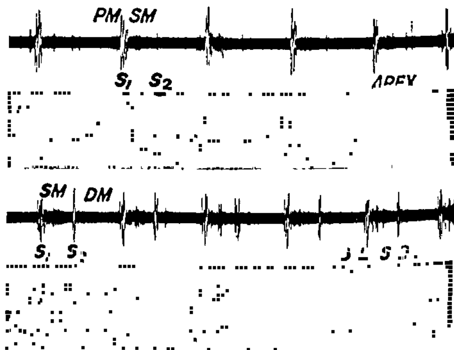


FIG. [illegible] sufficiency. Note systolic (SM), grade II, and short presystolic (PM), grade II, at apex (upper tracing). Lower tracing shows systolic murmur (SM), grade IV, and grade III early, blowing diastolic from the aortic valve.

does not mean that the patient has significant aortic stenosis. This patient had as his predominant lesion that of aortic insufficiency, and had a grade IV to V diastolic murmur along his left sternal border, which was associated with the low diastolic blood pressure reading characteristic of the severe forms of aortic insufficiency. No significant aortic stenosis was present, even though a grade IV aortic systolic murmur was heard.

The diagnosis of rheumatic aortic stenosis with slight aortic insufficiency and concomitant tight mitral stenosis was made in a 51 year old man (Fig. 357) only on the basis of a total clinical evaluation. He was referred for possible surgery for aortic stenosis, which was

evident by the typical harsh systolic murmur heard best along the left sternal border and aortic area In addition, a faint aortic diastolic murmur was present. There was no definite rumble at the apex, but the history of unusual dyspnea with the slightest exertion together with an enlarged left atrium that was noted on barium swallow and an enlarged pulmonary artery segment, suggested concomitant mitral stenosis. In addition the patient was fibrillating, which is unusual in

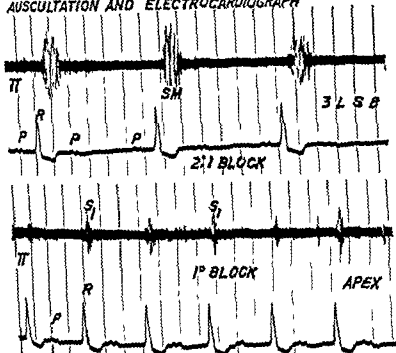
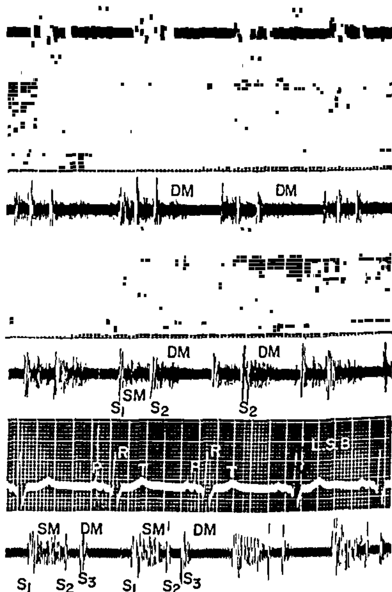


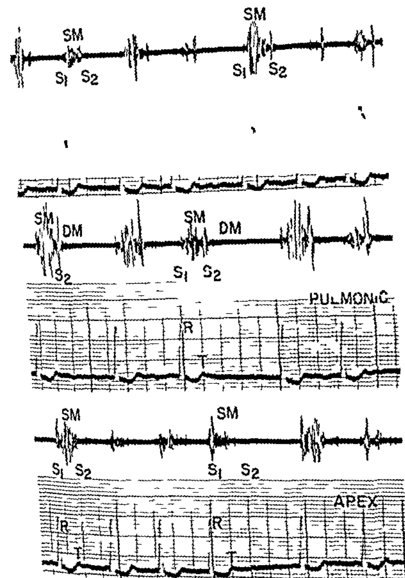
FIG. 355 Same case as Fig 175 Upper tracing shows diamond shaped murmur of aortic stenosis (SM) In lower tracing taken at the apex the first sound was accentuated in spite of prolongation of the P–R interval (0 36 second) Also note notching of the P waves From the combination of the auscultatory findings and electrocardiographic evidence the additional diagnosis of mitral stenosis was made clinically in spite of no significant diastolic murmur Postmortem examination revealed stenosis of both aortic and mitral valves

uncomplicated aortic valvular disease Left heart catheterization was performed and showed a significant gradient across the mitral valve, and at operation a mitral valve orifice the size of a lead pencil was found

Features of both aortic and mitral valve involvement are often readily evident and when combined with a total clinical evaluation a correct diagnosis may be made This was illustrated in a case of a 25 year old woman who had aortic stenosis and insufficiency plus



later th
gallop.
for aort
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ead pencil. ... mitral valve orifice found to be a ze

nitral stenosis and insufficiency (Fig 358) Her complaints were mainly related to the stenosis of the mitral valve It was felt that she had a relatively tight mitral stenosis, and associated aortic valve disease was evident from the presence of a grade IV harsh aortic systolic murmur and a grade III early, blowing diastolic murmur along

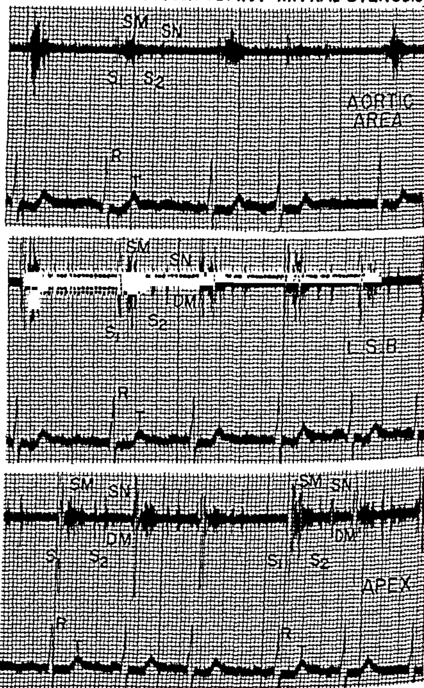
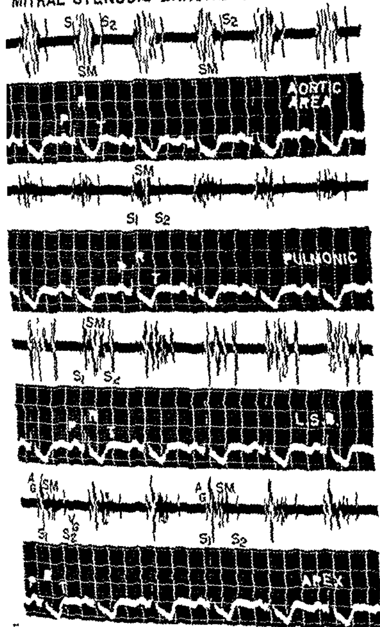


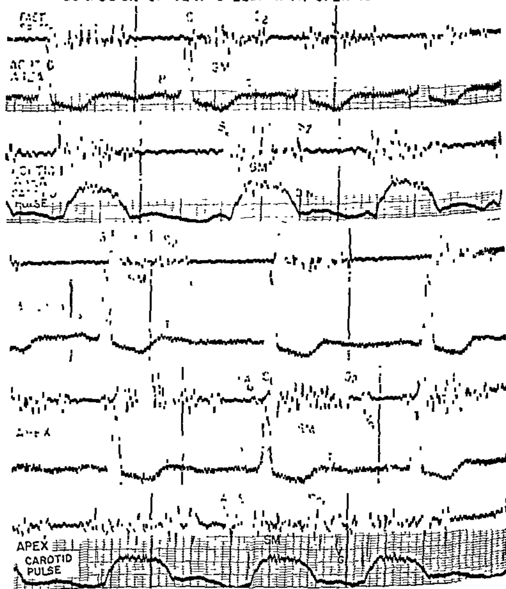
FIG. 360. A 50 year old woman with mitral stenosis plus aortic stenosis and insufficiency. Had grade IV harsh systolic murmur (SM) transmitted over precordium. Also grade III early, blowing diastolic murmur (DM) heard best left sternal border. At apex, note loud first sound (S_1), opening snap (SN) and diastolic rumble (DM). Atrial fibrillation present. Had mitral commissurotomy. No significant mitral insufficiency.



See also Fig 36?)

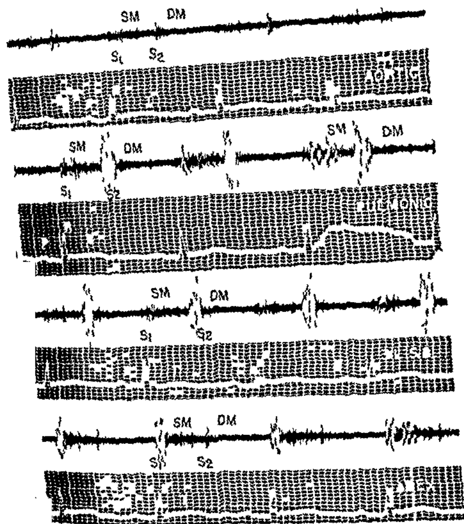
the left sternal border. At the apex the first sound was loud and a faint systolic murmur was present. An opening snap of the mitral valve was noted, and a diastolic rumble varying between grades III and IV followed the opening snap. Before a mitral commissurotomy was performed, the electrocardiogram showed atrial fibrillation, which was further support for a diagnosis of significant mitral valve involvement. At operation, an appreciable stenosis of the mitral valve was found, and this was opened. An aortic commissurotomy was not per-

AORTIC STENOSIS PROVEN AT AUTOPSY — S_2 DELAYED CAUSING CONFUSION OF VENT GALLOP WITH OPENING SNAP



ciency. Mitral valve was normal.

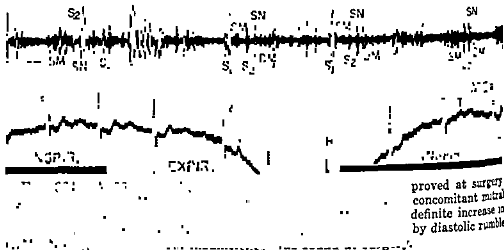
RHEUMATIC AORTIC AND MITRAL INSUFFICIENCY
(CONFUSED BY SOME c̄ PAT DUCTUS – CARD CATH NORMAL)



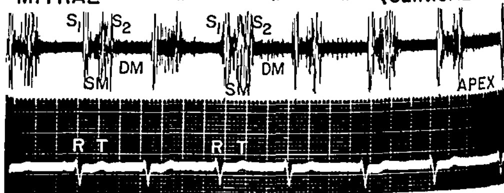
formed Figures 359 and 360 illustrate further examples of such combined lesions

Figure 361 When first seen this patient demonstrated classical findings of aortic stenosis with a minimal aortic insufficiency The diagnosis of concomitant mitral stenosis was made subsequently on the basis of hearing what appeared to be the opening snap of mitral ste

nosis and a diastolic rumble. In retrospect, a subsequent tracing (Fig. 362) illustrates why this erroneous diagnosis of mitral stenosis was made. Closure of the aortic valve was delayed, and, at the apex, a ventricular gallop rhythm associated with the patient's advanced heart failure was present. Because of this delay in the second sound,



proved at surgery concomitant mitral definite increase in by diastolic rumble



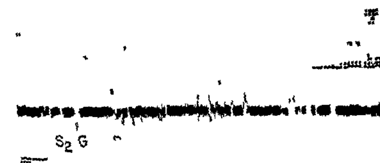
tolerance of severe congestive failure for a long period, lying flat without difficulty, venous engorgement, cardiomegaly, ascites and edema. No postmortem report obtained.

the interval between this and the gallop had the timing of an opening snap. In addition, a definite diastolic rumble, grade II-to-III, was evident at the apex, which at times may be present in aortic stenosis, particularly in instances of failure. Four years later, the patient had severe failure and her aortic systolic murmur had decreased several grades in intensity, causing some observers even to doubt the significance of this murmur and to question the [illegible] ity of aor[illegible]

nosis. As has previously been discussed in the presence of failure, murmurs such as those with aortic stenosis may decrease or even disappear at times. The patient died suddenly, and a postmortem examination revealed no involvement of the mitral valve—only a very severe stenosis of the aortic valve

As a rule the combination of mild to moderate aortic insufficiency with mild to moderate insufficiency of the mitral valve is not difficult





Fi... mitral valvular lesions Had pre... dominan... erately severe mitral stenosis (established post mor... ed (upper tracing) had systolic (SM) and diastolic (DM) murmurs ... note presystolic murmur (PM) and third sound (G) later in timing than opening snap of uncomplicated tight mitral stenosis. P waves notched on electrocardiogram.

to diagnose. However the combination of these lesions at times has caused *confusion* as illustrated in Figure 363 This 19 year old boy had murmurs over the pulmonic area and along the left sternal border that resembled a *continuous* murmur to such a degree that some observers made a diagnosis of patent ductus As shown on the phonocardiogram and as was *likewise* evident clinically this murmur did not envelop the second heart sound and therefore was atypical of patent ductus. However the *murmur* had a continuous quality and, after a complete cardiovascular work up, including cardiac catheter

ization, no evidence of a congenital heart lesion was present. It was felt that this patient had combined rheumatic mitral insufficiency and aortic insufficiency.

The combination of aortic insufficiency and mitral stenosis and the differentiation of the Austin Flint murmur have previously been discussed.

The combination of mitral stenosis and functional tricuspid insufficiency has also been discussed. Figure 364 represents a patient who

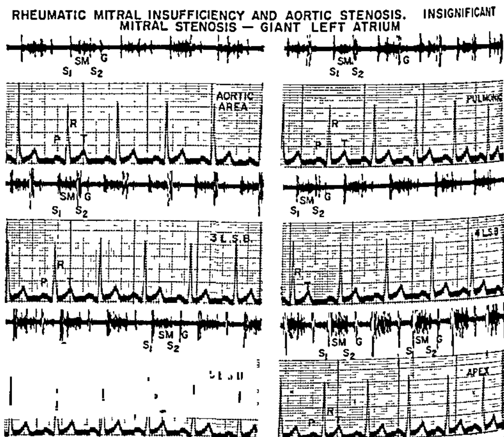


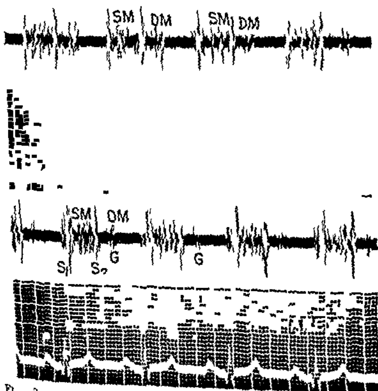
FIG. 367. Same patient as in Fig. 2
systolic murmur (SM). Along left ster
loud, pansystolic of different quality,
Loud sound in diastole (G) (ventricular
apical thrust. Had giant left atrium, also suggesting severe mitral insufficiency.

had mitral stenosis. However, coincident with inspiration there was an increase in the systolic murmur, and the possibility of a functional tricuspid insufficiency masquerading as mitral insufficiency arose. Left and right heart catheterization confirmed the presence of a tight mitral valve, and this was opened surgically. The possibility of tricuspid stenosis also existed, but the patient had not shown clinical features supporting that diagnosis, and tricuspid stenosis was not found on catheterization. The diastolic murmur may, as has been pointed out under Tricuspid Stenosis, increase coincident with inspiration; but likewise, in cases with tricuspid insufficiency and mitral stenosis, the rumble of mitral stenosis sometimes in-

eases coincident with inspiration, possibly due to increase in heart ate

Tricuspid stenosis in the majority of cases is of rheumatic etiology, lmost always is associated with mitral stenosis and frequently with ortic stenosis as well. The diastolic murmur of tricuspid stenosis as the same quality as that of mitral stenosis and its origin is nearer he midline Inasmuch as tricuspid stenosis almost always occurs in

RHEUMATIC MITRAL AND AORTIC INSUFFICIENCY– SIMULATED PAT DUCTUS



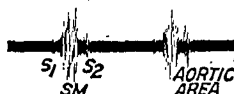
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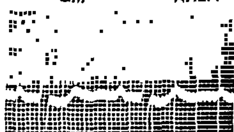
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patients who have mitral stenosis the possibility of tricuspid stenosis should be kept in mind and a search made Even the opening snap may be present in both conditions and as already discussed the possibility that this may be heard better at the tricuspid area may be misleading at times The patient whose case is illustrated in Figure 36a was examined regularly for many years She had long standing congestive heart failure and ascites and also demonstrated clinical signs of tricuspid stenosis and insufficiency At the apex

the first heart sound was accentuated, and there was a harsh systolic murmur that might well have been transmitted from her aortic stenotic murmur. A diastolic rumble was present, but a diagnosis of tricuspid stenosis was made after an evaluation of the total clinical picture. This patient tolerated obvious severe congestive heart

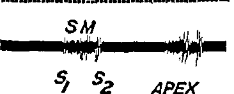
MITRAL STENOSIS & INSUFFICIENCY, AORTIC STENOSIS



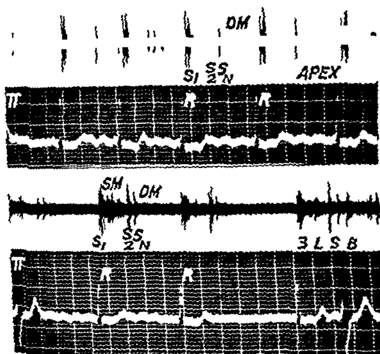




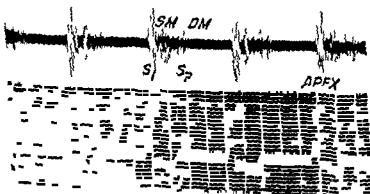






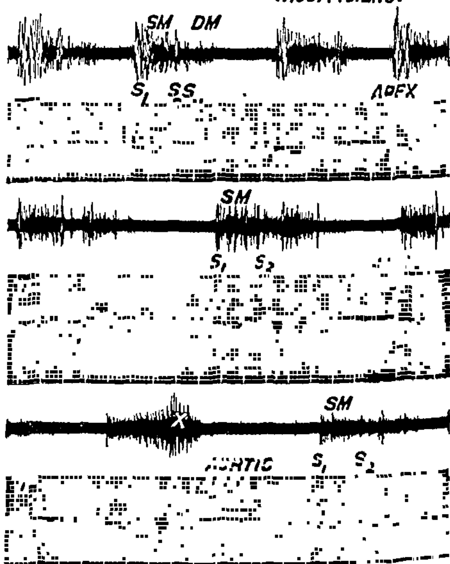


and insufficiency mitr



almost always associated

failure for a long time, and could lie flat without difficulty. She showed signs of venous engorgement, cardiomegaly, ascites and edema, however, and subsequently died. No postmortem report was obtained.



Other examples of combined aortic and mitral valve lesions are shown in Figures 366 through 372.

CONGENITAL HEART DISEASE

The importance of the diagnosis of congenital heart disease has changed tremendously in recent years. In the past it was mainly a

matter of academic interest whether a patient had a congenital defect of the heart, or what the exact abnormality might be No particular therapy was then effective, and only palliative treatments, such as bleeding, oxygen administration and general hygienic measures, were available Now that surgery offers the hope of cure in some conditions and amelioration in others, and since antibiotic therapy is very beneficial in some of the infectious complications, it has become imperative to make accurate anatomic diagnoses of congenital abnormalities of the heart.

It has been rewarding to find that, by using the total clinical evaluation of a patient, the great majority of uncomplicated congenital heart defects, particularly in the adult, can be diagnosed or suspected with a high degree of accuracy This kind of evaluation may be made by any physician, even in his office, and includes a careful and detailed history a physical examination an electrocardiogram, fluoroscopic and x ray examinations, and routine laboratory tests Some of the defects can be readily recognized even by simple bedside examination Others cannot at present be correctly appraised without specialized laboratory procedures, such as oxygen or dye determinations of the blood angiocardiography and catheterization studies of the heart These troublesome and involved methods of analysis are still absolutely necessary in many cases which cannot be solved in any other way but are not carried out unless the physician suspects the presence of congenital heart disease

The first suspicion that a patient has a congenital abnormality of the heart will generally be provoked by the detection of a cardiac murmur or by the finding of unexplained cyanosis and its sequelae clubbed fingers and polycythemia Less commonly an x ray examination will give the first clue At other times an unexplained hypertension or abnormal electrocardiogram may serve this purpose It is fair to say however that in most cases the interpretation of murmurs will be the initial step that may eventually lead to the diagnosis and for that reason auscultation of the heart is of vital importance It must be appreciated, however, that auscultation though occasionally sufficient in itself, is generally only the first step and must be aided by a complete study of the individual patient The following discussions therefore, will stress primarily the findings obtained with the stethoscope with special emphasis on the more common types of congenital heart disease seen in general practice

PATENT DUCTUS ARTERIOSUS

From the point of view of the practitioner the diagnosis of patent ductus is of primary importance because the condition is curable and generally not difficult to recognize The diagnosis should be made early in life, for the surgical mortality rate is much lower then than in older individuals, now having reached the low figure of 1 per cent

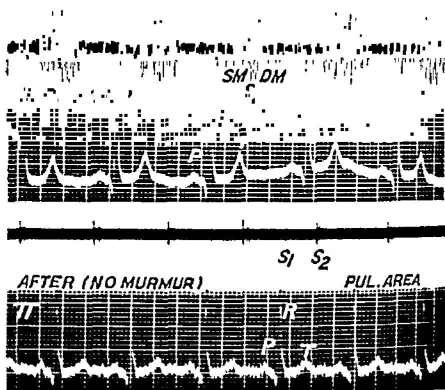


FIG. [illegible]
operat[illegible]
area. [illegible]
compo[illegible]

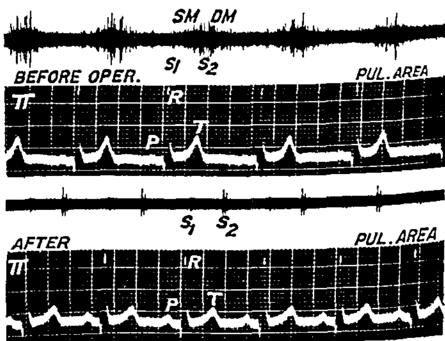
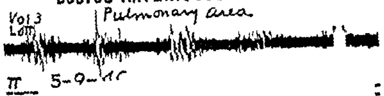


FIG. 374. A 19 year old girl. Upper tracing: Grade III continuous murmur (SM, DM) with accentuated late systolic component. Note peak of murmur is at second sound (S_2). Lower tracing, 11 days following operation; only grade I pulmonic systolic remained.

less for patients under 20 years of age Furthermore, in some in-
ances after the age of 30 the operation becomes more hazardous
d sometimes is even impossible to perform

In an uncomplicated case of patent ductus there often is very little
at one can detect of an abnormal nature apart from the character-
tic auscultatory findings There is no cyanosis or clubbing and many
tients ha e no cardiac enlargement. There is no significant electro-
ardiographic abnormality, and most patients have no dyspnea or
ardiac symptoms A few patients have stunted bodily development



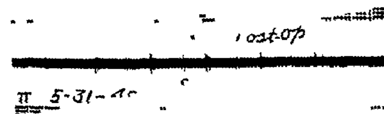




FIG. Girl age 7 had typical machinery type continuous murmur of patent uctus (upper tracing) which enveloped second sound (S) Following operation ower tracing) no murmur was pre ent.

r some evidence of cardiac weakness The pulse pressure is charac-
eristically increased, with a somewhat lowered diastolic level that is
omparable to readings obtained in patients with aortic insufficiency
The x ray findings are often helpful but are not always diagnostic
he most distinctive being a prominent pulmonary artery segment and
n exaggerated contraction of the left ventricle and great vessels In
all patent ductus patients there is a threat of the development of
bacterial endocarditis which used to be the cause of death in 25 per
cent of cases before the era of specific antibiotic therapy Formerly
the average age at death was only 35 years

MURMUR OF PATENT DUCTUS—BEFORE& AFTER OPERATION

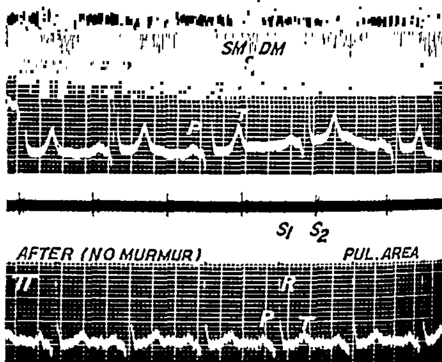


FIG [illegible]
opera [illegible]
area. [illegible]
compo [illegible]

"MACHINERY" MURMUR OF PATENT DUCTUS ARTERIOSUS BEFORE AND AFTER OPERATION

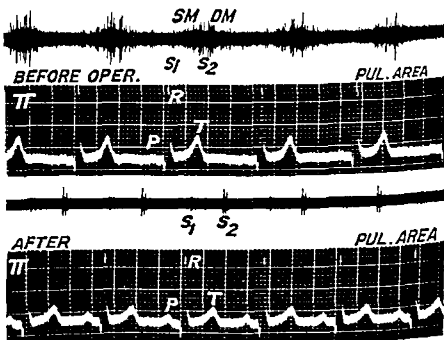


FIG. 374. A 19 year old girl. Upper tracing: Grade III continuous murmur (SM, DM) with accentuated late systolic component. Note peak of murmur is at second sound (S_2). Lower tracing, 11 days following operation; only grade I pulmonic systolic remained.

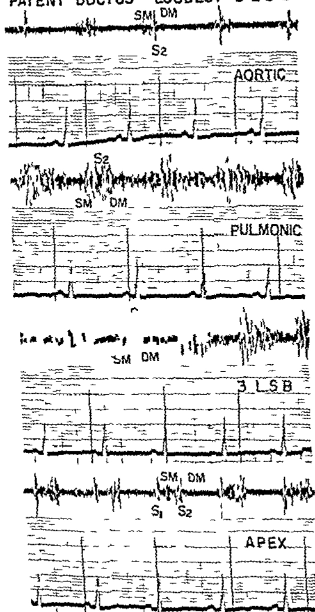


FIG. 377 A 20 year old man with typical continuous machinery murmur of patent ductus arteriosus. The murmur (SM–DM) enveloped the second sound (S_2) and was slightly louder at the third left sternal border (third strip) but almost as loud in the pulmonic area (*second strip*). Had successful surgical correction.

valve. The effect of respiration on the continuous murmur is variable. In some the murmur increases with inspiration (Fig 380) while in others it decreases (Fig 381). In many cases the murmur of patent ductus itself is sufficiently loud and characteristic to make the clinical

Auscultatory Findings. The murmur of patent ductus is commonly defined as machinery and continuous in character, and is heard best in the second left interspace (pulmonary area) (Figs. 373 through 376). Occasionally it is louder or equally loud in the third left interspace (Fig. 377). It appears to increase in intensity in the latter part of systole, envelop the second sound, and then continue throughout most or all of diastole. Characteristically, as the murmur envelops the second sound, the accentuation of the diastolic component is greatest in early diastole and gradually lessens toward

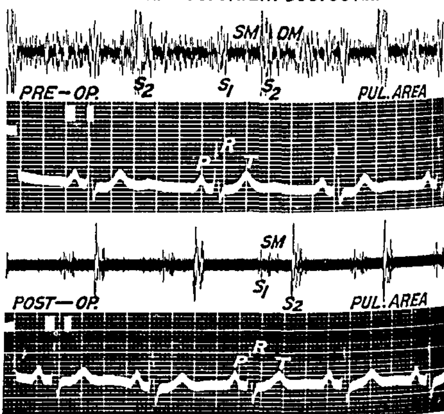
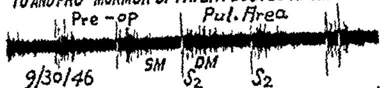
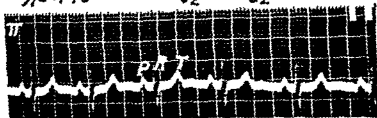


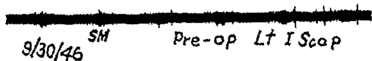
FIG. 376. Woman, age 22, with grade IV continuous machinery murmur (SM, DM) loudest over pulmonic area (upper tracing). Postoperatively (lower tracing) a grade I pulmonic systolic murmur (SM) was present.

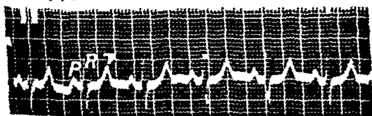
the end of diastole. It is generally louder in systole, but the reverse does occur (Fig. 378). When the murmur is very loud, as it often is, it can be heard all over the chest (Fig. 231), and even in the back (Fig. 232). Although in the typical case the basal murmur is continuous, in some it has a to-and-fro quality (Fig. 379) with a clear systolic and diastolic component, simulating acquired aortic valvular disease. Not infrequently one hears a faint diastolic murmur at the apex that suggests mitral stenosis. This apparently is due to the very rapid ventricular filling that occurs with this condition and the increased flow of shunted blood to the left heart across the mitral

"TO AND FRO" MURMUR OF PATENT DUCTUS ARTERIOSUS



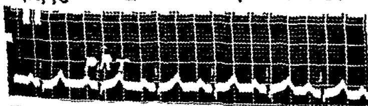






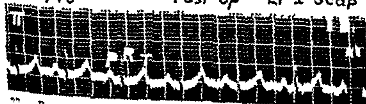
Ductus Sectioned 10/8/46





S S_2 S_1 S_2

10/17/46 Post-op Lt I Scap



diagnosis certain. However, with increasing experience, we have learned that patent ductus may be present with very inconspicuous findings. Sometimes the murmur is faint, of grade II or III intensity (Figs. 382, 383).

The most important auscultatory point in diagnosis is a continuous murmur that envelops the second heart sound. When the second sound is not so enveloped by a murmur that at first might be suspected to be that of patent ductus, one should consider the possibility that he may be dealing with another lesion that simulates patent ductus, or that

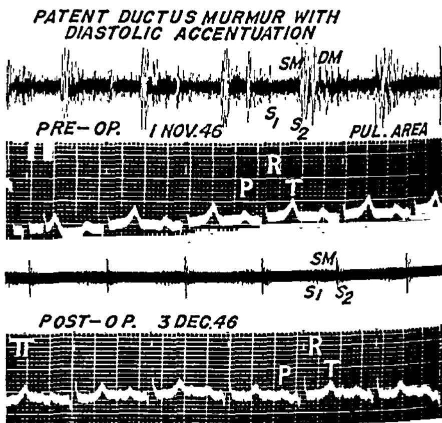
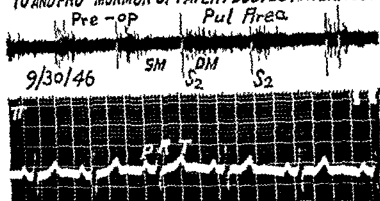


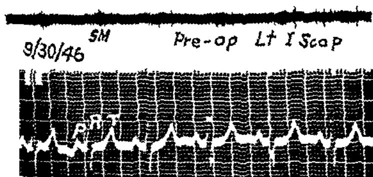
FIG. 378. Man, 35 years of age. Had grade III machinery murmur (upper tracing) with loud diastolic component (DM) and fainter systolic (SM). After operation (lower tracing), a grade II pulmonic systolic murmur (SM) persisted.

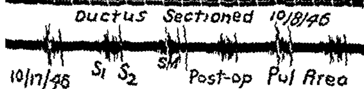
an associated defect may be present. When an associated congenital defect is present the typical continuous machinery murmur often changes, combining the auscultatory features of both. At other times the murmur of one lesion may predominate.

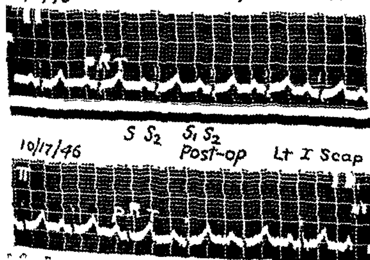
An example of coarctation in addition to patent ductus is shown in Figure 384. A classical ductus murmur was absent. Instead, a systolic murmur with maximal intensity in midsystole and a faint diastolic component was heard. The phonocardiogram of a four year old girl with patent ductus and pulmonic stenosis is shown in Figure 385. A grade V continuous murmur was loudest over the pulmonic area and was well transmitted over the entire precordium. Auscultation could

"TO AND FRO" MURMUR OF PATENT DUCTUS ARTERIOSUS



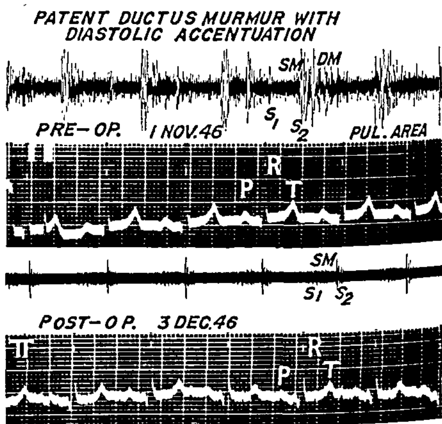






diagnosis certain. However, with increasing experience, we have learned that patent ductus may be present with very inconspicuous findings. Sometimes the murmur is faint, of grade II or III intensity (Figs. 382, 383).

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not establish the diagnosis of concomitant pulmonic stenosis in this child, but proof of right ventricular hypertrophy by x ray and/or electrocardiographic examination would furnish such a clue It is important also to listen carefully with both the bell and the diaphragm of the stethoscope. Sometimes the murmur is heard best with the diaphragm and at other times it is heard better using the bell with light pressure Although it is not certain, there may be a correlation between the size of the ductus and the sound frequency of the mur-

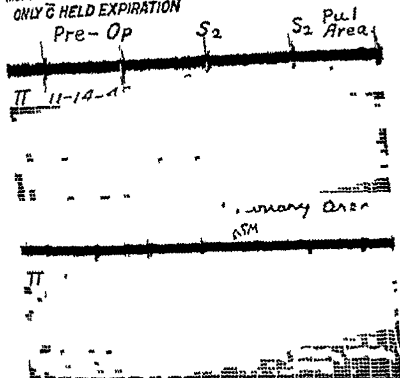


Fig. ... years old with a grade II systolic murmur (SM) and a ... grade I continuous diastolic component (DM) heard only in held expiration (upper tracing) Following operation (lower tracing) only a grade I systolic murmur was heard in the pulmonic area

mur There appears to be some evidence that a small ductus may have a murmur of higher frequency and that it may be the one heard best with the diaphragm of the stethoscope The lower frequencies that are heard best with the bell may be related to the larger ductus However there appears to be no clinical correlation between the size of the ductus and the intensity of the *murmur* We have had instances in which the systolic component was rather faint or the diastolic entirely inaudible. In others no murmur whatsoever was evident, and rarely a loud, early, blowing diastolic murmur of insufficiency of th

CONTINUOUS MURMUR OF PATENT DUCTUS INCREASING $\bar{c}$ INSPIRATION

SM DM

SM DM

Fig. 380. Woman, age 36, with patent ductus arteriosus proved at surgery. With inspiration there was a striking increase in intensity of the continuous murmur (SM–DM).

MURMUR OF PATENT DUCTUS DECREASING $\bar{c}$ INSPIRATION

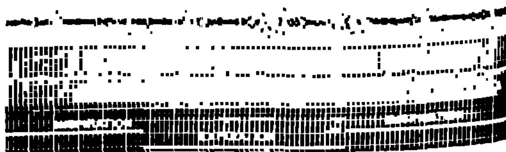


Fig. 381. Typical continuous machinery murmur (SM–DM) [illegible] sound (S_2) in an 8 year old boy. The murmur decreased on inspiration. Had successful surgery.

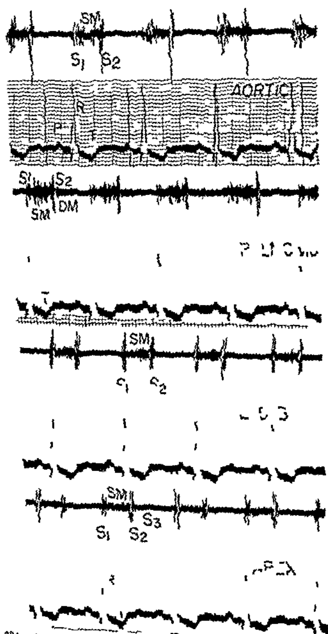
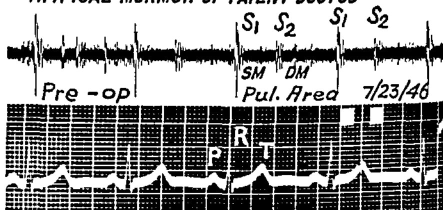
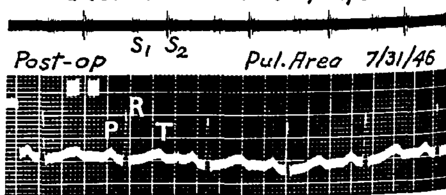


FIG. 234. Girl, age 19 with patent ductus arteriosus and coarctation of the aorta. Murmur was not typical of patent ductus arteriosus. Had a louder systolic murmur (SM) with maximum intensity in midsystole. In addition, a faint diastolic component (DM) heard best over pulmonic area (second strip). Both defects corrected at surgery.

pulmonary valve would be evident that at first might even be confused with aortic insufficiency.

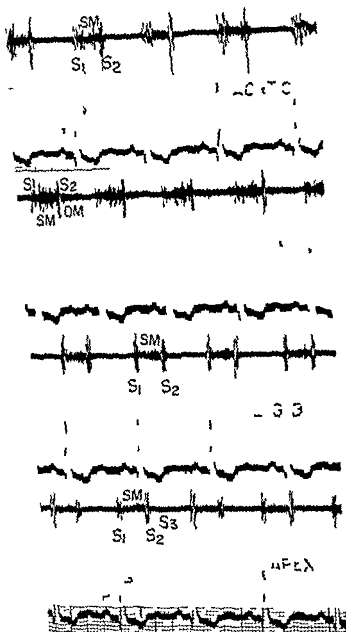
These changes in the typical auscultatory findings are commonly associated with the development of associated pulmonary hypertension. At a previous examination carried out a year or so before these phonocardiograms were made, we regarded at least one of these patients as having no heart disease whatsoever, because only a slight pulmonary systolic murmur was heard. No doubt there are many





ation.

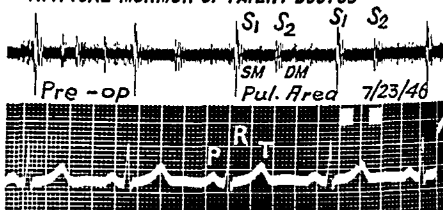
other such cases that are diagnosed as a "benign systolic murmur." It is sometimes difficult to avoid these errors, unless by chance an x-ray examination reveals some suspicious abnormalities. The only means available to establish a definite diagnosis in such atypical cases is catheterization studies, which can reveal unequivocal findings in the majority of cases. If the maximum intensity of the murmur is in the aortic rather than the pulmonary area, one should suspect that the ductus comes off a right aortic arch. However, other possibilities, such as perforation of the *sinus of Valsalva*, truncus arteriosus and pulmonary a-v fistula, should also be considered. The findings of pat-

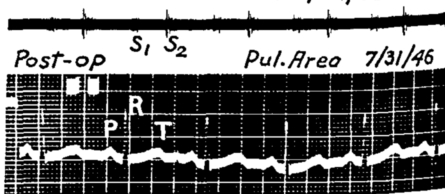


(DM) heard best over pulmonic area (second strip) Both defects corrected at surgery

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not rule out the possibility of patent ductus on the basis of an absence of the continuous murmur until the first few years of life have passed This delay is not significant, as the operation would probably not be performed for a few years even if the diagnosis were clear, except in very rare instances when an operation may become imperative even in the first few months of life In very early life, congenital defects may be multiple, and should patent ductus be one of them an operation is not likely to be indicated

After a successful operation the murmurs entirely disappear, or, as occurs in many cases a slight pulmonary systolic murmur may persist. The latter is due either to a dilatation of the pulmonary artery that may persist for some weeks months or even years or to the same much debated mechanism that underlies other functional pulmonic systolic murmurs Figure 386 387 and 388 show pre- and post operative auscultatory findings of a 33 year old woman Cardiac and pulmonary artery enlargement were present and an unusual feature was a greatly enlarged left atrium At operation a large ductus was sectioned The pulmonic and apical systolic murmurs that were present several weeks after operation (Fig 387) became progressively fainter after nine months (Fig 388) If a definite basal diastolic murmur remains after surgery it would indicate that there is an accompanying lesion, such as aortic insufficiency which might have developed as part of a bacterial endocarditis or that restoration of the patent ductus has occurred The latter is always a possibility when the duct is ligated and not sectioned However we have observed one patient who after a successful section of her ductus, had a persistent grade II diastolic blowing murmur presumably caused by an insufficiency of the pulmonic valve associated with dilatation of the pulmonary artery

Patent Ductus with Associated Pulmonary Hypertension It is reasonable to state that 90 to 95 per cent of patients with patent ductus arteriosus have a characteristic machinery type of murmur If one does not hear this murmur in the adult or child (after the first several years of life) the possibility of patent ductus is most unlikely As illustrated by the case of a 20 year old woman in Figure 389 an unnecessary operation for a mistaken diagnosis of patent ductus can be prevented by making sure that a continuous murmur is present. However the development or association of [illegible] nsion [illegible] egree [illegible] stolic [illegible] 390, 391) there may be no murmur (Fig 392) or occasionally a fainter "to-and fro" type (Fig 393), or sometimes only a diastolic murmur The *diastolic* murmur on occasions may be high

ent ductus become quite complicated if there are other congenital anomalies, but fortunately those cases that are most suitable for surgical intervention generally will have *no other defects.*

During the first few years of life the signs of patent ductus may

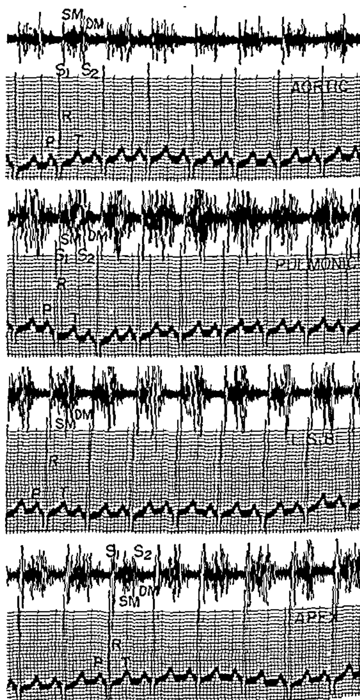


FIG. 385. *A 4 year old girl with patent ductus plus pulmonic* stenosis. Had grade V continuous murmur (SM–DM) loudest over pulmonic area and well *transmitted over entire precordium.* From auscultation alone the diagnosis of pulmonic stenosis could not be made in this case. Had cardiac catheterization and surgery.

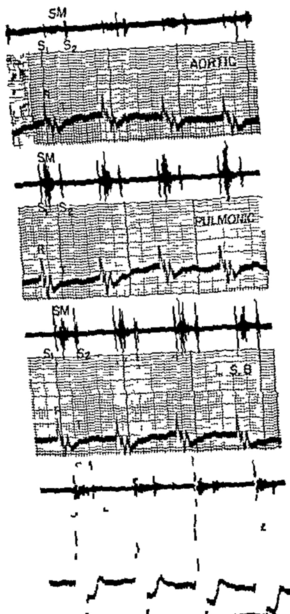
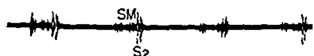


Fig. 387 Same patient as Fig 386 Continuous murmur absent, but systolic murmur (SM) and left bundle branch block persist several weeks after operation

pitched and blowing (Figs 394 395) suggesting incompetence of the pulmonary valve In such a case it generally follows an accentuated closely split second pulmonic sound At times a systolic murmur or diastolic rumble (or both) are heard at the apex In some cases the continuous murmur is intermittent and its presence and dis-

PATENT DUCTUS—BEFORE OPERATION







3 4 LSB



APEX

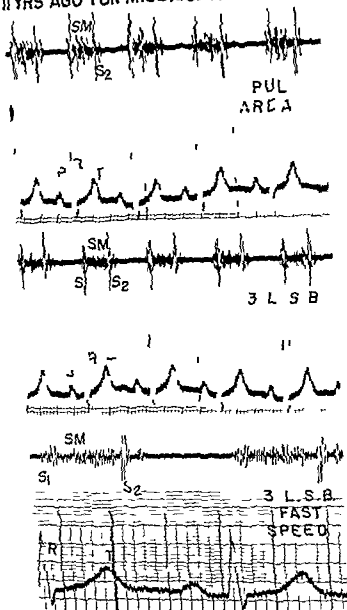
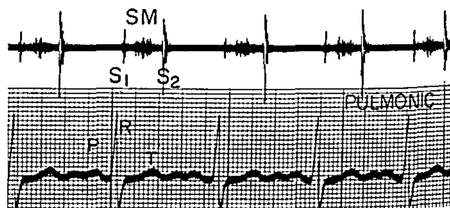


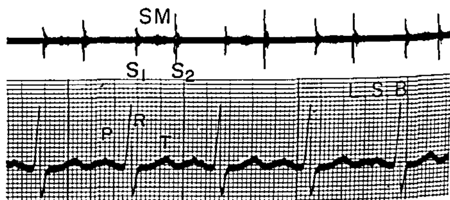
FIG. 389 Part A Woman 20 years of age who had operation 11 years *prior for sus*

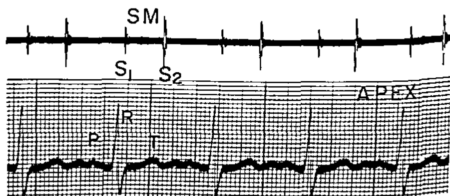
appearance suggest a fluctuation of the level of pulmonary hypertension.

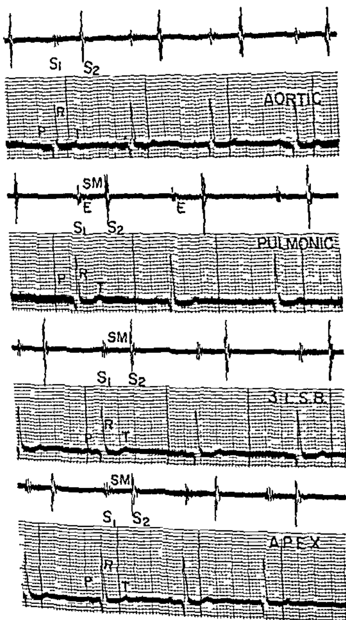
Cyanosis may be absent, slight or marked, and its degree is dependent on the degree of reversal of flow through the ductus. When present, cyanosis may be limited to the lower extremities, or may be

PATENT DUCTUS – 9 MOS. POST-OPER.








PATENT DUCTUS c̄ PUL. HYPERTENSION
S1
S2
AORTIC
R
P
T
SM
E
E
S1
S2
PULMONIC
P
R
T
SM
S1
S2
3 L.S.B.
R
P
T
SM
S1
S2
APEX
R
P
T

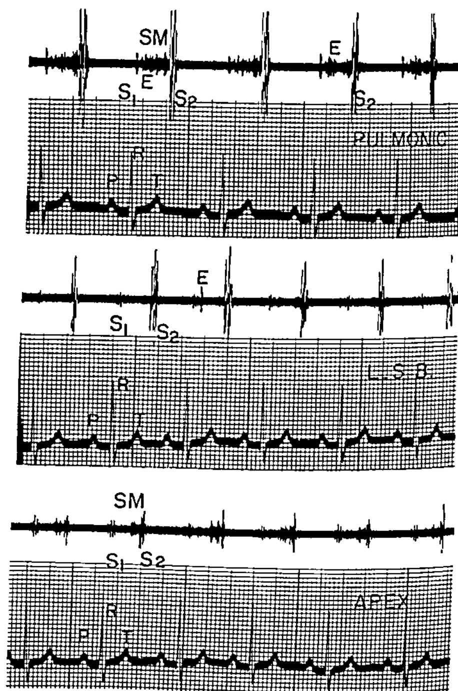


FIG. 390. A 6 year old girl with patent ductus and pulmonary hypertension with

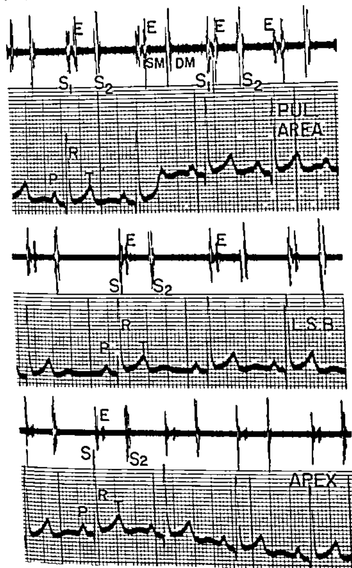


FIG 93 A 28 year old woman with patent ductus pulmonary hypertension and reversal of flow Had striking difference in cyanosis of toes as compared with fingers. No typical continuous patent ductus murmur Instead had faint grade II systolic (SM) and diastolic (DM) murmurs The second sound (S_2) over the pulmonic area was very loud and closely split. A prominent ejection sound (E) was heard loudest over pulmonic area but transmitted over entire precordium

be paradoxical becoming narrower with inspiration As a rule, the second heart sound can be heard best by listening along the middle left sternal edge.

With the development of pulmonary hypertension however the second heart sound (Figs 390 391 393–395) characteristically becomes progressively more accentuated and more closely split In fact

greater there than in the upper (*Figs. 390, 391, 393, 394, 395*). This discrepancy, together with an absence of the typical machinery murmur, may be the first clue in suspecting this condition, as was the case with the patient illustrated in Figure 390 who had previously been diagnosed as having an Eisenmenger complex.

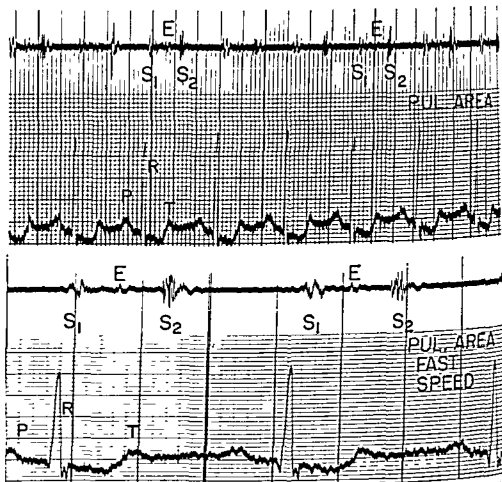
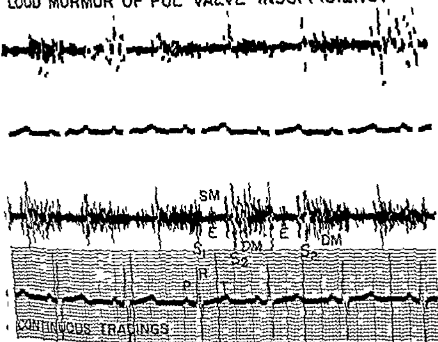


FIG. 392. A 30 year old woman with proven patent ductus arteriosus and pulmonary hypertension. Had a balanced shunt, as demonstrated on cardiac catheterization where catheter pas
sure were balanced with a
study, based on finding of
femoral artery drawn simultaneously. Had no significant murmurs. An ejection sound (E) was noted in approximately midsystole and was heard best over the pulmonic area.

HEART SOUNDS. In uncomplicated patent ductus the heart sounds are of little diagnostic importance. As a rule the machinery murmur enveloping the second sound obscures the second sound over the pulmonary area. In other cases where the murmur is less intense, the second heart sound may be of normal intensity or accentuated, and is normally split (Fig. 396). In some instances the splitting is noted to

n a number of our patients who have had pulmonary hypertension ith reversal of flow and atypical murmurs of patent ductus, the iagnosis was first suspected by the interpretation of a loud, accen-

nstances, the patient should be asked to perform an exercise that



might accentuate the cyanosis of the toes One of our recent patients (Fig 390) *had a very accentuated second so nd*

... with the first ... that led to a correct diagnosis

A *pulmonary systolic* ejection sound is common in the presence of pulmonary hypertension (Figs 390 392 393 395) The electrocardiagram in the *uncomplicated* case is usually normal With *the de*velopment of pulmonary hypertension however right ventricular hy

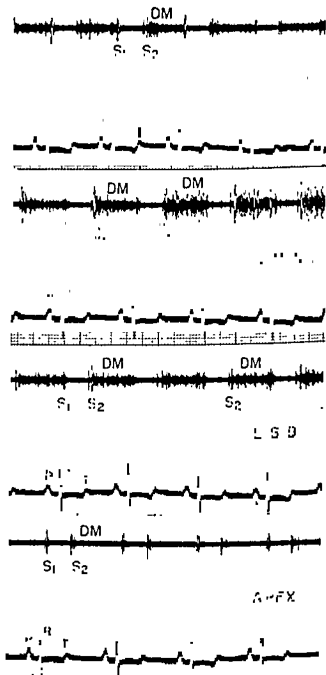
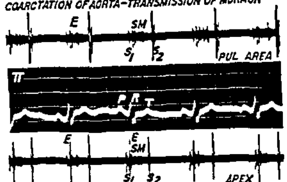
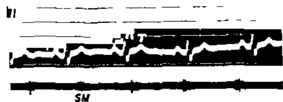
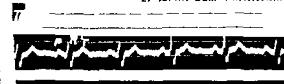
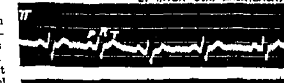


FIG. 394. Woman with patent ductus and pulmonary hypertension proved on cardiac catheterization. No typical machinery murmur of patent ductus. Instead, had loud blowing diastolic murmur of insufficiency of pulmonary valve (DM) heard best over pulmonic area (second strip), but also heard well over aortic area and along left sternal border.

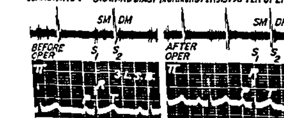












…G. 397 First case Man …ears old with coarctation …orta, resected Note ejec… sound (E) heard over base … apex. A grade III systolic …rmur (SM) was present at … base (first tracing) grade …pical systolic (SM) (second …cing) grade II systolic …M) in left posterior cervical …angle (third tracing) and no …rmur in left interscapular …on (fourth tracing) At the …ght base of chest posteriorly …fth tracing) definite collat…al pulsations of intercostal …teries were felt and a grade … systolic murmur (SM) was …ard over the vessel

Second case (lowest trac…ig). A 2 year old man with …oarctation of aorta and prob…ble bicuspid aortic valves …aortic systolic (SM) and di…astolic (DM) murmurs present …before and after successful …operation.

pertrophy may be evident. Fluoroscopic and x-ray examinations in the uncomplicated case may show enlargement of the main pulmonary artery and its branches, varying from slight to moderate. The vascular markings likewise vary from normal to increased. As a rule, the heart size is normal, but occasionally some enlargement is found. There is often left atrial enlargement, presumably due to the increase in the amount of blood carried by the left side of the heart. With the association of pulmonary hypertension, the pulmonary artery segment and its branch may show further enlargement, peripheral lung fields generally become less vascular, and an enlargement of the right ventricle takes place.

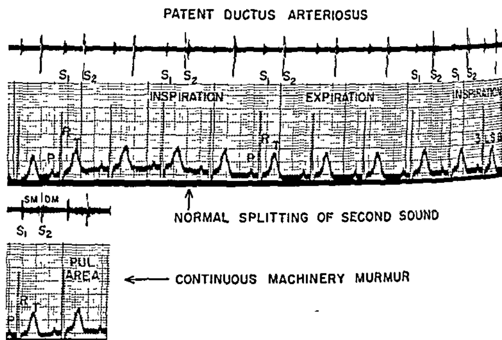


FIG. 396. A 4½ year old boy with typical findings of patent ductus arteriosus corrected at surgery. Continuous murmur with late systolic (SM) and early diastolic (DM) accentuation which enveloped second sound (S_2) (lower tracing). Had normal splitting of second sound (upper tracing) widening with inspiration and single with expiration.

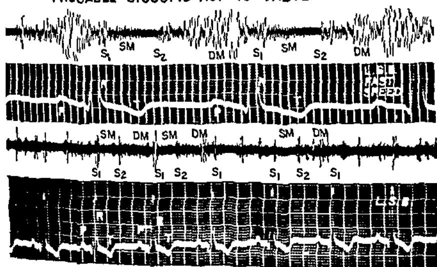
In patients who have a persistent reversal of flow through a ductus caused by pulmonary hypertension, surgical closure of the ductus has caused death in the great majority of cases (Fig. 390).

COARCTATION OF THE AORTA

With the development of surgical methods for the cure of coarctation of the aorta, it has become important to recognize this condition and to do so in the early years of life. The operation has already been successful in many cases, but the results are much more promising when performed upon patients under 30 years of age. Coarctation of the aorta is present in about one in 3000 to 5000 of our population, so it is not extremely rare. It is also of interest that bicuspid aortic

ical examination hypertension in the arms and a basal systolic murmur It is, therefore, imperative to think about the possibility of coarctation of the aorta in all cases of *hypertension*, especially in *individuals* With this in mind, it will be helpful to develop the habit of feeling for pulsations in the femoral arteries and abdominal aorta as a routine procedure in all hypertensive cases In patients with coarctation the systolic blood pressure is elevated in the arms and not in the legs, whereas in others the level is greater in the legs This peculiarity is readily overlooked as it is not customary to take readings of the legs A clue that this should be done is afforded when arterial pulsations are absent or only feebly felt below

COARCTATION OF AORTA – SEVERE AORTIC INSUFFICIENCY
PROBABLE BICUSPID AORTIC VALVE

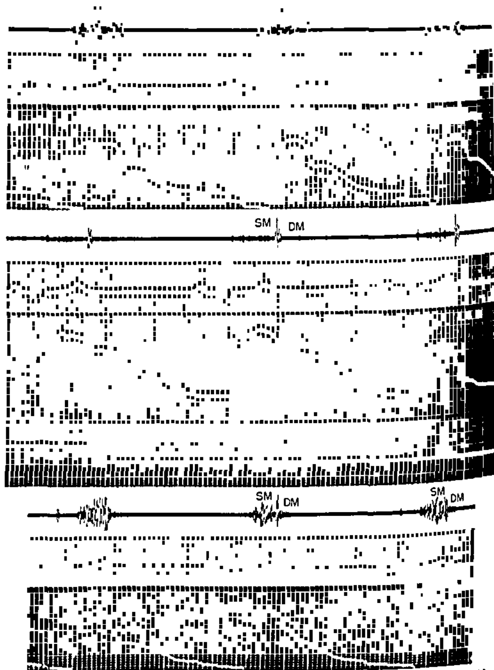


the chest. When the diagnosis is suspected, further observations can be made Careful palpation over the back of the chest may reveal prominent pulsating arteries which are *not present normally* An x ray examination of the chest may show notching of the lower edges of the ribs which is practically pathognomonic or absence of the aortic knob on the left In this way very accurate preoperative diagnoses can be made

The second simple finding is a *basal systolic murmur* heard best in the pulmonary and/or aortic areas (Figs 225 397) This of itself would be of little value as basal systolic murmurs are quite common in a variety of types of heart disease and even in patients without organic heart disease What is more helpful is that a systolic murmur is audible *in* the interscapular region or over some of the large

valves (Figs. 397, lowest tracing, 398, 399, 400, 401, lowest tracing) and congenital cerebral aneurysms are frequently associated with coarctation of the aorta.

The diagnosis will generally be initiated on the basis of one or the other of two abnormal findings that can be elicited on routine phys-



left scapular area, back (lower strip) a murmur was present late ... and extended into early diastole (DM).

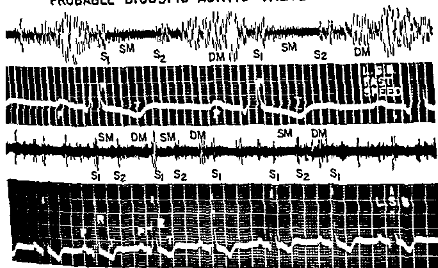
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and [illegible] readily overlooked as it is not customary to take



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arteries of the back. However, it has been made clear that any very loud cardiac murmur may be transmitted to the back. What is particularly characteristic of coarctation is that the systolic murmur can be clearly heard in the back even when anteriorly it is only slight or

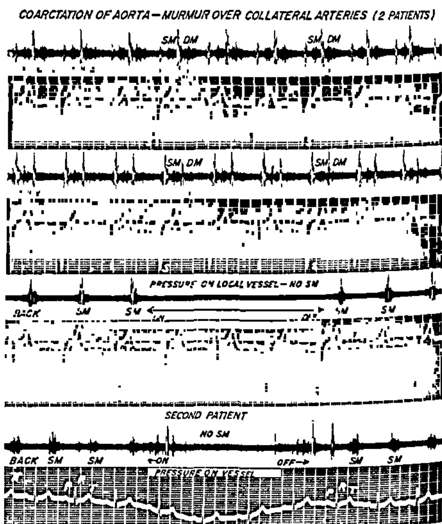
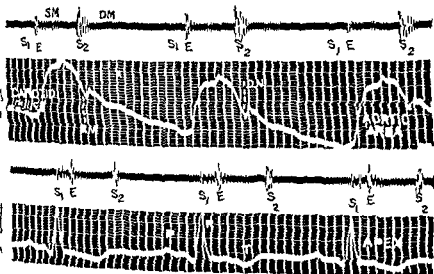


FIG. 400. … insufficiency. … base. Also gr… apex. A grad… peared on local pressure (see … was suspected because of di… right arm 210/74; right leg u… classic coarctation of aorta. … sel of back on local occlusion of artery Operation successful. In both patients, the murmur (SM) heard over the back (lower two tracings) appeared to extend into early diastole.

moderate in intensity (grade II or III). It generally is less intense posteriorly but may be as loud. The reason for this is that the place of origin of the murmur, for the most part, is about as far from the front as the back of the thoracic cage. Another mechanism to explain a dorsal systolic murmur is the enlarged tortuous compensatory ar-

teries that are frequently present. Some murmurs are made locally in these vessels This explains both the presence of a systolic murmur even when the coarctation is complete, and its persistence in some cases after a successful operation In the first case illustrated by Figure 397 there were several auscultatory points of interest. The murmur was heard particularly well high up in the chest, in the left supraclavicular region in the left suprascapular region and in the area Furthermore, a systolic murmur was audible over a prominent vessel below the angle of the right scapula and was louder than the murmur over the left lower scapula This observation helped to confirm the fact that the murmur was not transmitted from the



aorta but was made locally In this case there was also a faint but definite aortic diastolic murmur at the left sternal border an occasional finding in cases of coarctation of the aorta

Observations on the two cases shown in Figure 400 afford definite proof that some of the systolic murmurs ...

... temporarily following local digital occlusion of the arteries.

A systolic murmur generally one grade louder than over the base or back is almost always heard over the supraclavicular fossae Presumably this *murmur is also produced by the* engorged collaterals As

recorded in Figures 400 and 407, the systolic murmur over the back appears late in systole and extends into early diastole. This is apparently due to a lag between the actual systole of the heart and the resultant flow of blood through the collaterals or through the coarcted area.

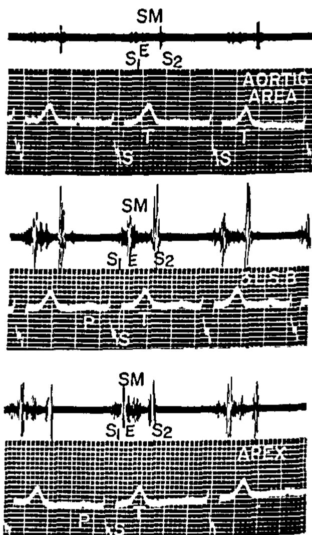


FIG. 402. A 7 year old patient with coarctation of aorta, ventricular septal defect and pulmonary hypertension. Systolic murmur (SM) was louder along left sternal border and apex than at base. An ejection sound (E) was heard best at apex (lower strip) and also along left sternal border and aortic area.

Associated defects should be considered when additional murmurs are heard over the precordium. A continuous machinery murmur at the pulmonic area suggests patent ductus, although such a sound can be caused by coarctation without associated patent ductus, as illustrated in Figure 403. On the other hand, the absence of the typical

of patent ductus does not rule out that condition as a possible for in some patients the diastolic component is not heard possible explanation for this latter situation in some patients with coarctation and patent ductus is the development of pulmonary hypertension When the degree of pulmonary hypertension approaches that of systemic pressure the diastolic component of the ductus murmur becomes fainter or disappears We recently studied two adult patients with proven combined coarctation and patent ductus Both were admitted to the hospital for surgery for coarctation, and at that time neither was suspected of having patent ductus *One patient had* only the usual systolic murmurs of coarctation and no diastolic murmur the other had a prominent grade IV systolic *murmur* with only a *faint* diastolic component *(Fig* 404)

A combination of ventricular septal defect and coarctation of the

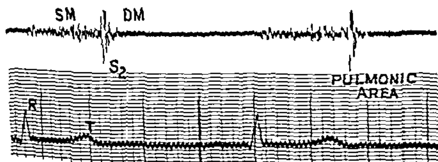
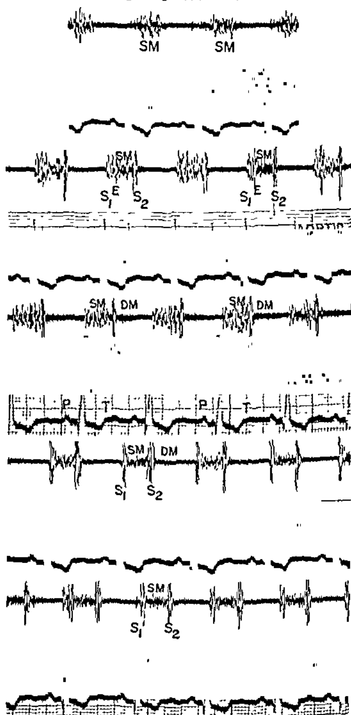


Fig. 403. Girl age 11 with coarctation of aorta. Had continuous murmur (SM-DM) heard over pulmonic area and back Patent ductus suspected but not found at operation.

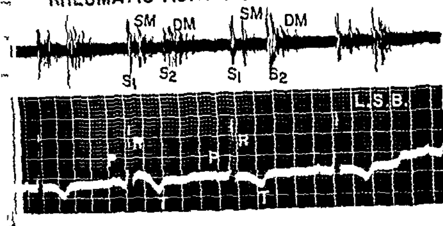
aorta is shown in Figure 402 The presence of a louder murmur that is heard best along the lower left sternal border particularly if it is associated with a palpable thrill over this area would suggest this associated defect. Additional *findings of enlargement of the pulmonary* artery increased vascularity of the lungs and an *electrocardiogram* showing any evidence of right ventricular hypertrophy would rule out an uncomplicated coarctation

Louder systolic murmurs alone or in combination with diastolic murmurs should lead one to suspect associated defects A grade IV or louder basal systolic murmur necessitates ruling out aortic or subaortic stenosis This would be particularly true in a patient with a loud rough systolic *murmur* over the aortic area and a palpable thrill that radiates *toward the right neck* or shoulder An early blowing high pitched diastolic murmur heard over the aortic area and/or along the left sternal border (Figs 388 through 401) *is consistent* with a diagnosis of aortic insufficiency from incompetent

bicuspid aortic valves. Such a murmur may, however, represent an associated rheumatic aortic insufficiency, as demonstrated in the post-mortem examination of the patient illustrated in Figure 405. Occasionally, even severe aortic insufficiency is produced by an incom-



F
sys
(thi...
mur (SM) was easily heard.



tion was mild The patient was first to receive Hufnagel valve. Died approximately four years later

COARCTATION OF AORTA — EJECTION SOUND AT AORTIC & APICAL AREAS

S1 E S2 S1 E S2 E

AORTIC AREA

S1 E S2 S1 E S2

APEX

FIG. 406. A 15 year old boy with coarctation of the aorta later corrected by surgery Had distinct early systolic ejection sound (E) at apex and aortic areas.

petent bicuspid valve, and the patient with such a condition seeks medical advice because of the finding of a diastolic murmur, congestive heart failure, or both. In such an individual a loud, early, blowing diastolic murmur (e.g., grade IV) may be transmitted along the left sternal border and to the apex. A murmur of relative mitral stenosis (Austin Flint type) may likewise be present. Such cases are most commonly diagnosed as of rheumatic etiology. The presence of coarctation is overlooked because the femoral vessels in such patients are often easily palpated, and in fact, may be accentuated. However,

COARCTATION OF AORTA — PRE-OPERATIVE

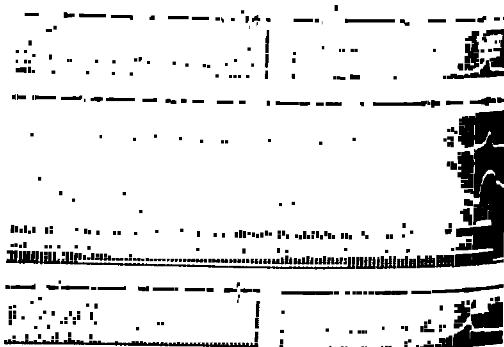


FIG. 407. Boy, age 16, had typical findings of coarctation. At apex (upper left) note systolic murmur (SM) and early ejection sound (E). Over the pulmonic area (middle tracing) note systolic murmur (SM) and normal splitting of second sound sound (S_2).

the radial pulsations are still stronger than the femorals when both are compared simultaneously. This discrepancy may be confirmed by blood pressure determinations in the arms and legs.

Figure 405 illustrates such a combination. This 31 year old woman had the severe form of aortic insufficiency resulting in progressive symptoms of congestive heart failure. At surgery, where a Hufnagel valve was inserted in an attempt to relieve the leak of the aortic valve, a surprise finding was that of associated coarctation. This was not recognized preoperatively because the femoral pulsations were excellent. In retrospect, the diagnosis might have been suspected if more attention had been paid to the slightly lower systolic blood pressure in

lower extremities, as compared with the upper In addition, a systolic murmur, unusual for the average case of aortic insufficiency, was well heard in the interscapular area This woman was the very first patient to have the Hufnagel valve operation and this experience ... the possible significance of the above findings and ...

After her operation this patient having done ... developed an aneurysm at the site of insertion of the plastic valve The valve had been placed at a point just beyond the long, mildly coarcted segment (technically the only area at the time of operation that would fit the valve) It is known that the area distal to a coarcted

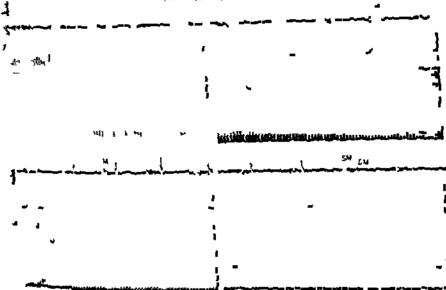


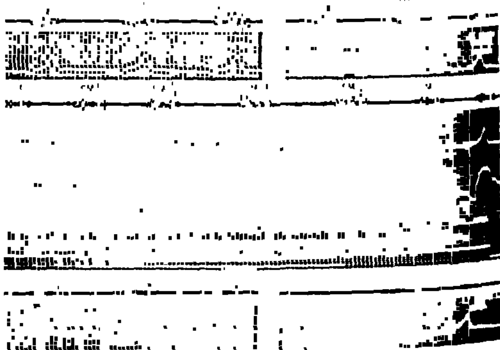
Fig 408. Same patient as Fig 407 Note persistence of murmurs and ejection sound (E) after surgery

area is prone to be weak or friable and this contributed to the formation of the aneurysm in this patient. She died as a result of her second operation

tion If such a sound is present, mitral stenosis is suspected particularly if the diastolic rumble is associated with other auscultatory evidence such as an opening snap and a loud *first heart sound*

Heart Sounds with Coarctation An early systolic ejection sound is generally heard over the aortic and mitral areas (Figs 397, 398 401, 406) This is sometimes mistaken for the first heart sound The aortic component of the second sound may be accentuated and the

petent bicuspid valve, and the patient with such a condition seeks medical advice because of the finding of a diastolic murmur, congestive heart failure, or both. In such an individual a loud, early, blowing diastolic murmur (e.g., grade IV) may be transmitted along the left sternal border and to the apex. A murmur of relative mitral stenosis (Austin Flint type) may likewise be present. Such cases are most commonly diagnosed as of rheumatic etiology. The presence of coarctation is overlooked because the femoral vessels in such patients are often easily palpated, and in fact, may be accentuated. However,



sound (S2).

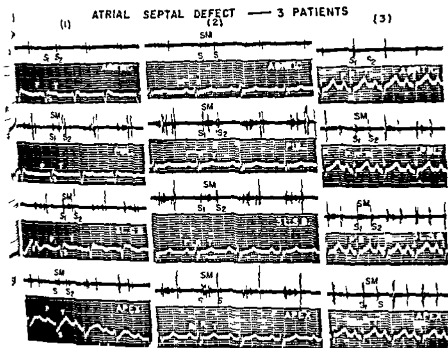
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this patient were identical to those of coarctation, which had been the preoperative diagnosis The obstruction caused by the band was readily relieved at surgery

ATRIAL (AURICULAR) SEPTAL DEFECT

Uncomplicated Atrial Defects (Ostium Secundum Type) Uncomplicated atrial defects usually can be diagnosed on clinical grounds. Since the predominant shunt is from left to right through the defect



in the atrial septum cyanosis is absent except in the complicated cases where associated defects or pulmonary hypertension are present. Blood flow is increased through the right atrium right ventricle pulmonary artery and the lungs with little of the burden placed on the left ventricle The right atrium is enlarged often greatly and there is concomitant enlargement of the right ventricle and the pulmonary artery

atrium is not

enlargement

greatly enlarged Right atrial and right ventricular enlargement in

which the patient suffered thrombosis of the aorta, the condition was diagnosed by aortography and was cured by surgery. There must be other such cases that have been caused by automobile accidents.

Congenital Band of the Aorta Simulating Coarctation. A congenital band occurring at the usual site of a coarctation was found at operation in the patient shown in Figure 410. The clinical features in

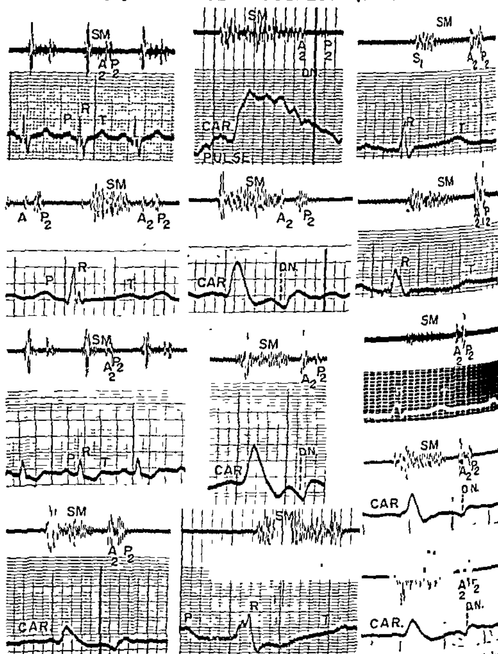


FIG. 411. Composite of 13 patients with atrial septal defect showing systolic murmur (SM) usually heard best over pulmonic area but occasionally loudest at third left sternal border. Note wide splitting of second sound (A_2P_2) characteristic of uncomplicated atrial defect.

he absence of an increase in size of the left atrium on barium swal ow examination plus vascular lung fields, make a roentgenologic picure that is frequently seen. The electrocardiogram will aid in the liagnosis by showing incomplete or complete right bundle branch ock, or right ventricular hypertrophy in the great majority of ases. The presence of marked right ventricular hypertrophy, how ver, should make one suspect either associated pulmonary hyperten n or another defect such as pulmonic stenosis or ventricular septal fect.

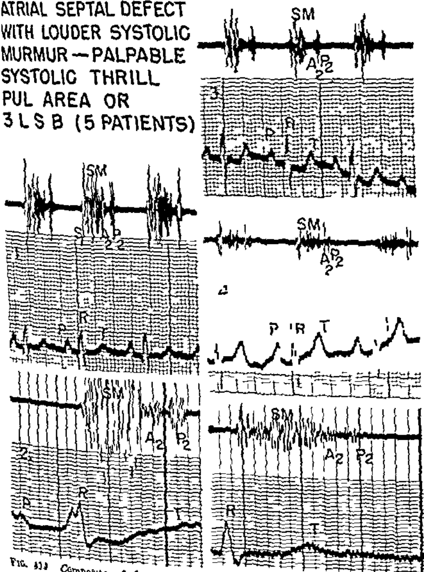
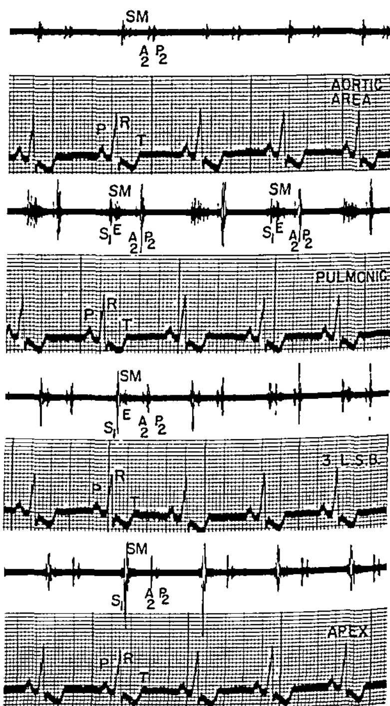


FIG. 311 Composite of five patients with proven atrial septal defect. Note systolic murmurs, grade IV or above and associated palpable systolic thrill.



[illegible] A 52 year old woman with proven atrial septal defect. Had wide [illegible]

[illegible]

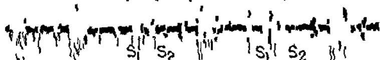
is also heard well over pulmonic area and left sternal border. At ap[illegible] tracing) first sound (S_1) moderately accentuated; varies with respiration.

early blowing

may be heard along the left stern

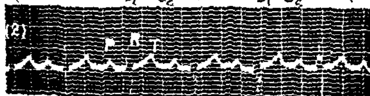
aphragm of the stethoscope is used (Fig 415) At the mitral or

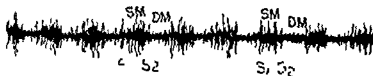
ATRIAL SEPTAL DEFECT c̄ SYST MURMUR
+ DIAST RUMBLE AT APEX– 3 PATIENTS

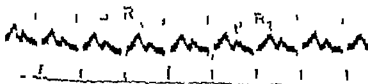












had an apical

sure

defect and cleft

to catheterization.

The auscultatory findings in atrial septal defect consist of a systolic murmur (average grade III) that is usually heard best over the pulmonic area (Figs. 411, 412, 413). Occasionally it is heard best at the third or fourth left intercostal space. The murmur begins early

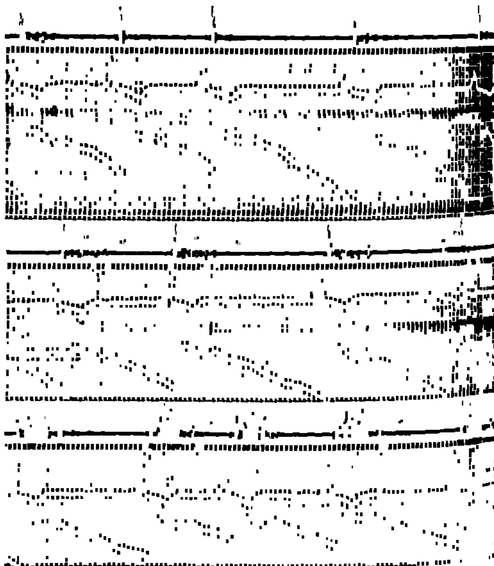
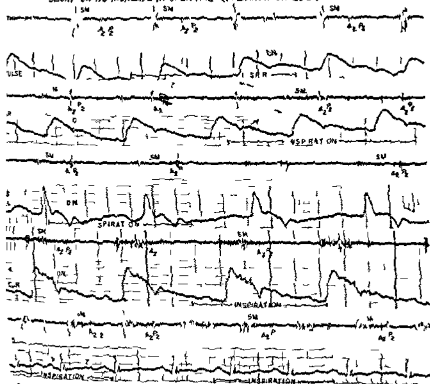


FIG. 415. A 38 year old woman with atrial septal defect. Had systolic murmur (SM) heard best over pulmonic area (lower tracing) and also an early, blowing diastolic murmur over pulmonary area and left sternal border (lower tracing). Note wide splitting of second heart sound (A_2P_2) (second tracing). At apex (upper tracing) first sound (S_1) slightly accentuated and faint systolic murmur (SM) present.

in systole, ending shortly after midsystole. As a rule the systolic murmur with atrial defect is not loud enough to be associated with a palpable thrill. Less commonly, however, the murmur is grade IV or louder and a thrill is present (Fig. 414). Diastolic murmurs are

whose second sound became single with expiration would represent a rare exception The second component of the second heart sound is the delayed closure of the pulmonary valve. Following surgical closure of the defect, however, splitting of the second sound generally became normal (Figs 421 422) We have observed the normal changes of splitting due to respiration even in instances where the electrocardiogram still demonstrated a right ventricular conduction delay after operation The intensity of the pulmonic component of the second

5 PATIENTS WITH ATRIAL SEPTAL DEFECT — WIDE SPLITTING OF 2nd SOUND INSPIRATION PRODUCES SLIGHT OR NO INCREASE IN SPLITTING (PUL AREA OR LSB)



sound varies and when accentuated, suggests increased pressure from pulmonary hypertension

An early systolic sound or ejection sound may be heard over the pulmonic area (Figs 113 422 423) and is possibly more common when pulmonary hypertension is associated with the defect. This sound is often misinterpreted as the first heart sound since it occurs early in systole As shown in Figure 422 one can easily see how such a misinterpretation can be made However by comparing the ejection sound with the apical first sound it will be noted that the

tricuspid-area a diastolic rumble may be present, usually in a well localized spot. This sound is detected better with the bell of the stethoscope (Fig. 416). With inspiration, the murmur may increase in intensity, suggesting that the rumble may be due to an increased flow across the tricuspid valve. At the same time, a systolic murmur is often present at the apex or at the tricuspid area, representing an

5 PATIENTS WITH ATRIAL SEPTAL DEFECT—WIDE SPLITTING OF 2nd SOUND—INSPIRATION PRODUCES SLIGHT OR NO INCREASE IN SPLITTING (PUL AREA OR L S B)



insufficiency of the tricuspid valve, or possibly of the mitral valve as well.

The auscultatory sounds are helpful in diagnosis. A wide splitting of the second heart sound is present in the pulmonic area (Figs. 411, 417, 418, 419), and this is an important point in the majority of patients with uncomplicated atrial septal defects. Some patients, however, have a wide splitting that increases further on inspiration (Fig. 420). The patient with uncomplicated ostium secundum atrial defect

normal or even diminished in intensity Probably the conflicting that influence the first sound are the increased blood flow the tricuspid valve and the conduction disturbance The former would tend to increase and the latter to decrease the intensity of the sound

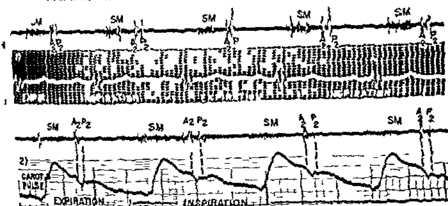


FIG. 4 0 Two patients with proven atrial septal defect showing wide splitting of second heart sound (A P_) over pulmonic area. The splitting increased slightly with inspiration in each but never became single on expiration Note systolic murmur (SM) in each.

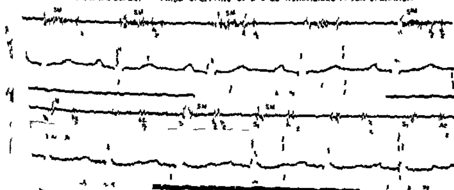


FIG. 4°1 Woman age 26 with typical systolic murmur (SM) and fixed splitting of second sound (A P₂) before operation (upper tracing) Following operation (lower tracing) note splitting of second sound (A_2P_2) becomes normal widening with inspiration and becoming close or single with expiration

Most patients with atrial defect experience difficulty early in life but occasionally a patient may live a normal life span This latter possibility was demonstrated by the case of a 67 year old woman (Fig. 423) whose enlarged pulmonary arteries were mistaken for lymphoma. Forty x ray treatments had previously been given over this "lymphoma.

ejection sound comes early in systole rather than coincident with the first heart sound. It is well to think of an enlarged pulmonary artery whenever the "first sound" over the pulmonic area is as loud or even louder than the second sound, for the second sound is generally louder over the aortic and pulmonic areas. Exceptions to this general rule are

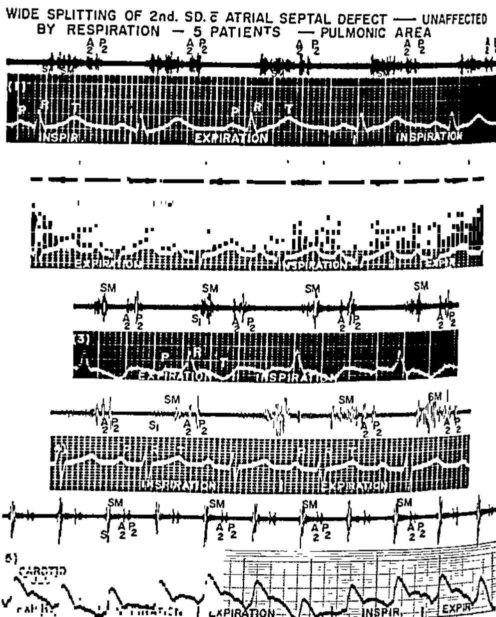


FIG 419. Five patients with atrial septal defect showing systolic murmur (SM) and wide fixed splitting of second sound (A_2P_2).

mitral stenosis and a short P–R interval, which produce such a loud first sound at the apex that it is also well heard at the base. However, if these exceptions have been ruled out, one can suspect that a loud "first sound" over the pulmonary area is actually an ejection sound. The first heart sound in atrial septal defect varies. In approximately one-half of these patients, the sound is accentuated; in the others it

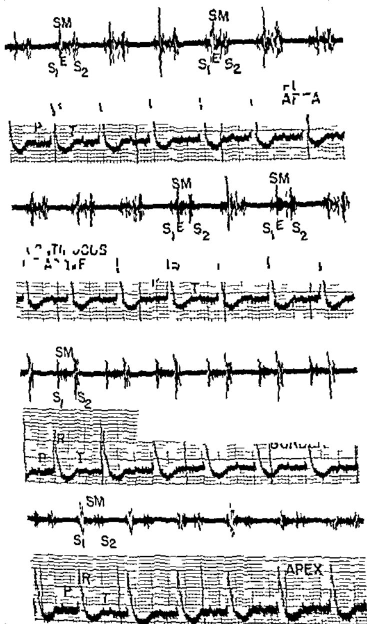
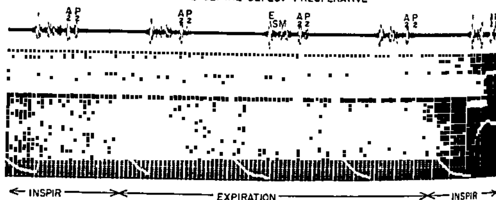
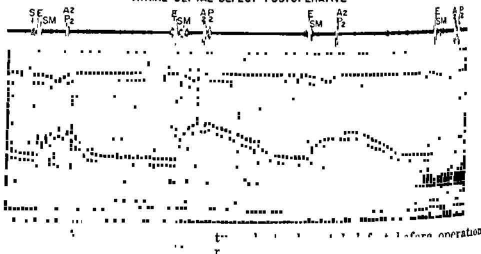


FIG. 4?3 67 year old woman with

Atrial Septal Defect Plus Pulmonary Hypertension. The physical findings in these patients vary, depending on the degree of pulmonary hypertension. The pulmonic component of the second heart sound becomes less widely split, increases in intensity, and is often easily palpated (Fig. 424). The systolic murmur is still present, and, in addition, an early, blowing diastolic murmur of insufficiency of the





[illegible] before operation (lower tracing) a faint systolic murm[illegible] splitting of second sound (A_2P_2) became normal, widening with inspiration and becoming single with expiration.

pulmonary valve is sometimes heard along the left sternal border. An apical systolic and diastolic rumble also may be present.

Surgical closure in the usual type of uncomplicated atrial septal defect (ostium secundum) can now be performed, and the mortality rate after such an operation is low. Because of this it is important that the diagnosis be established and an operation performed in selected patients before pulmonary hypertension has developed, or while it is in its early stages before irreversible changes have taken place. To determine this factor, however, cardiac catheterization studies will

...se of atrial septal defect the presence of a loud pulmonic systolic murmur, grade IV to VI, and a palpable thrill that is directed toward the left neck and shoulder region should suggest the possibility of associated pulmonic stenosis because the murmur of atrial septal

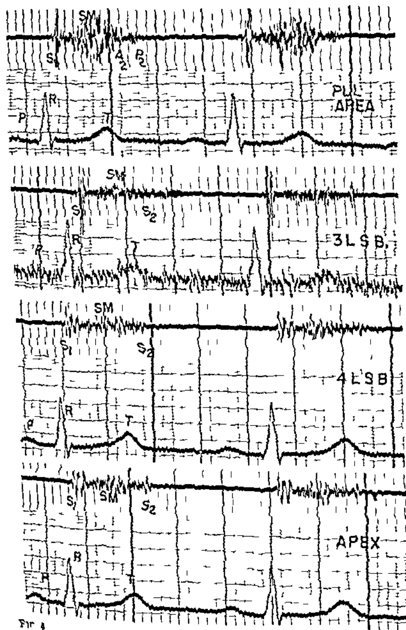
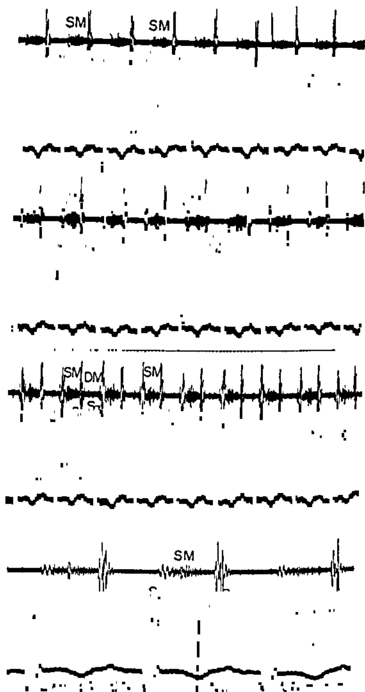


Fig 4

...onic stenosis ... was longer ... was delayed and faint. ... the second sound The pulmonic



be necessary. After surgery, the pulmonary systolic murmur and the ejection sound may persist, as illustrated in Figures 421 and 422. A postoperative decrease in the intensity of the murmur is common.

ATRIAL SEPTAL DEFECT PLUS PULMONIC STENOSIS. In a suspected

case of atrial septal defect the presence of a loud pulmonic systolic murmur, grade IV to VI, and a palpable thrill that is directed toward the left neck and shoulder region should suggest the possibility of associated pulmonic stenosis, because the murmur of atrial septal

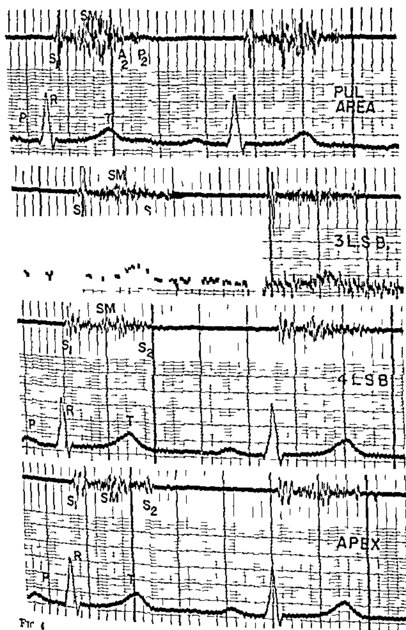


Fig 4

lmonic stenosis
M) was longer
The pulmonic

and faint.

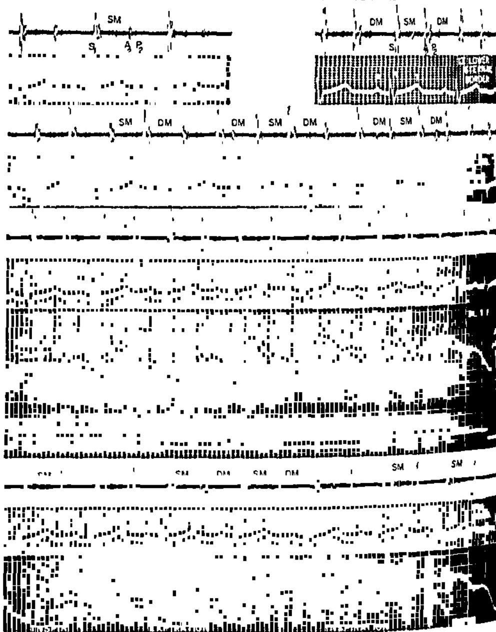


FIG 426. A 10 year old boy wit
cultatory findings of atrial defect
ting of second sound over pulmonar
was evident along the third left sternal border and left lower sternal border.
twin brother with atrial defect plus pulmonic stenosis. (Fig. 427).

defect is generally not loud enough to produce a palpable thrill. The characteristic wide splitting of uncomplicated atrial defect may now be altered so that over the pulmonic area only one component, the delayed pulmonary valve closure, is evident On careful auscultation the rough, harsh systolic murmur of pulmonic stenosis is heard (lasting longer than the customary atrial septal murmur), and extends through the aortic valve component of the second sound often mask-

ng it. At an interval after the murmur a distinct but diminished ound may be heard which represents the delayed pulmonary valve losure of the second sound (Fig 425) By listening for the splitting long the lower sternal border (third or fourth interspace) the murmur is *less evident* and a *wide splitting* is present In this area the tenotic murmur of the pulmonic valve does not mask the aortic valve losure of the second sound

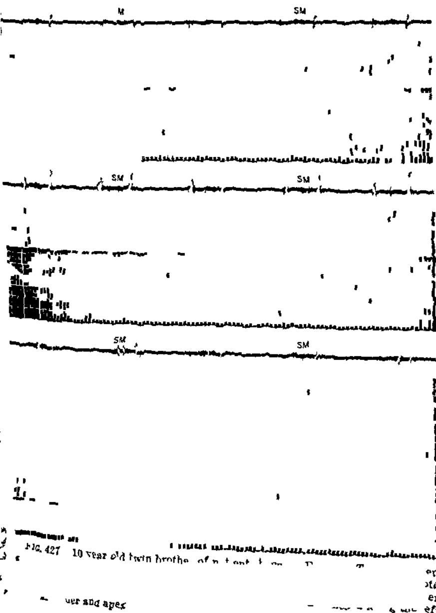
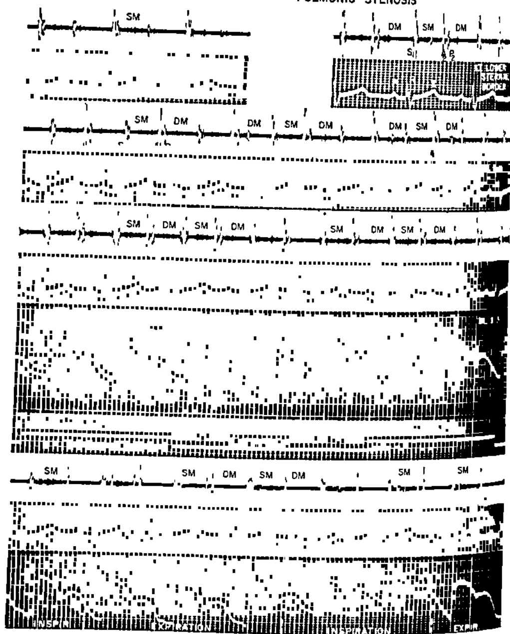
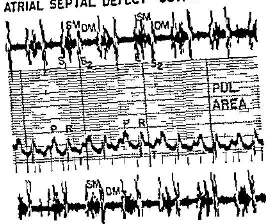


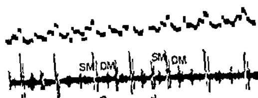
FIG. 427 10 year old twin brothe of p t ent ... en ... ote ... er ... uer and apex ... eft

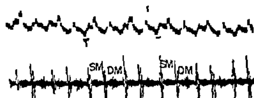


twin brother with atrial defect plus pulmonic stenosis. (Fig. 427).

defect is generally not loud enough to produce a palpable thrill. The characteristic wide splitting of uncomplicated atrial defect may now be altered so that over the pulmonic area only one component, the delayed pulmonary valve closure, is evident. On careful auscultation the rough, harsh systolic murmur of pulmonic stenosis is heard (lasting longer than the customary atrial septal murmur), and extends through the aortic valve component of the second sound often mask-





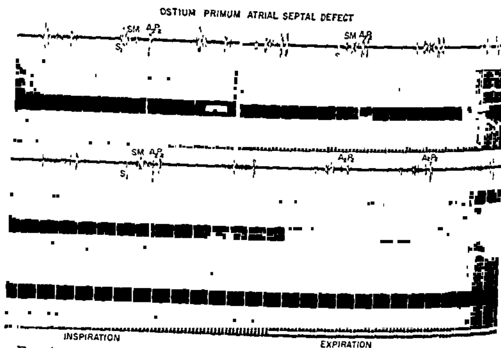




areas, including tricuspid area (bottom tracing) Had early systolic ejection soun (E) over pulmonary area.

The lung fields are apt to be less vascular and the electrocardiogram shows right ventricular hypertrophy if a significant pulmonary stenosis is present. This problem will be discussed further in the section on Congenital Pulmonic Stenosis.

ATRIAL SEPTAL DEFECT IN TWIN BROTHERS. Figures 426 and 427 show the auscultatory findings in twins, both of whom had atrial septal defect. In one, pulmonic stenosis was also present (the phonocardiogram in Figure 427 was made after this patient had had an operation for pulmonic stenosis). The typical wide, fixed splitting of the second sound was present in both patients, together with apical



... f second sound (A_2P_2) which was unaffected by respiration. Along lower left sternal border and apex, note systolic murmur (SM).

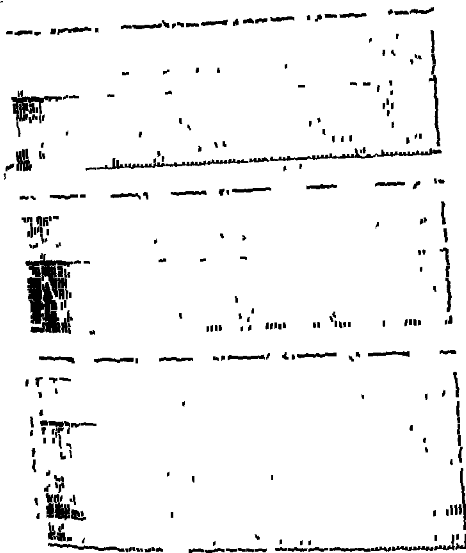
and pulmonic systolic murmurs. In one (Fig. 426), a diastolic murmur was well heard along the left sternal border.

Atrial Septal Defect (Ostium Primum Type). This type of atrial septal defect is less common than the ostium secundum that has already been discussed. However, it is important to differentiate the ostium primum type, if at all possible, because it presents a much more difficult technical problem (Figs. 428, 429). If the surgeon is aware that an ostium primum defect is present, he will not attempt an operation unless open heart surgery is available.

The ostium primum defect occurs low in the atrial septum just above the a-v valves, and, as a result, there may be a deformity of these valves. This deformity often manifests as mitral insufficiency.

Persistent Atrioventricular Canal (Persistent Ostium Atrioventriculare Commune)

Closely allied to the ostium primum and, in fact incorporating an ostium primum atrial defect, is the persistent atrioventricular canal in addition to the low atrial defect there is an incorporated defect of

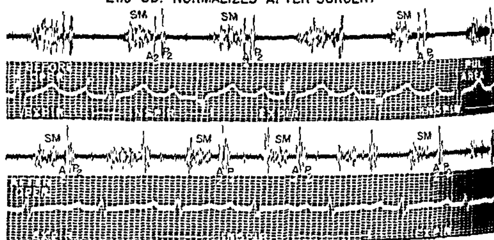


Boy age 13 with ostium primum defect and mongoloid features At subsequent operation atrial ... artially ... nd wide ... sternal ... murmur (SM) ... third sound (S_3) and atrial gallop ... terval 0 21 second.

The x-ray findings in such a patient may be quite similar to those of the more usual atrial septal defect, and auscultatory features likewise may be entirely similar (Fig. 428). One important differential clue is electrocardiographic evidence of left axis deviation. When this is present in a patient previously thought to have the usual atrial septal defect, ostium primum is the probable diagnosis.

However, the findings on auscultation may be different. Because of the associated mitral valve insufficiency, a systolic murmur is more frequently heard at the apex in patients with an ostium primum defect (Fig. 429). Over the pulmonary area, a systolic murmur, averaging grade III intensity, is usually heard. This is generally of short duration. The second sound may vary, depending on the presence or absence of pulmonary hypertension. In the absence of sig-

ATRIAL SEPTAL DEFECT—OSTIUM PRIMUM TYPE— WIDE FIXED SPLITTING OF 2nd SD. NORMALIZES AFTER SURGERY



(lower tracing) Note persistence of systolic murmur (SM) after surgery.

nificant pulmonary hypertension, the splitting of the second sound is wide, relatively fixed, and varies little or not at all with inspiration. It thus simulates the usual (ostium secundum) atrial defect. Figure 430 illustrates a patient with ostium primum defect both before and after operation. Before surgery (upper tracing), the second sound was widely split and showed no change with respiration. After corrective surgery (lower tracing), the splitting became normal.

If pulmonary hypertension is present, the second sound is accentuated, and the splitting is closer and may show a slight increase with inspiration. These changes are similar to those of the usual atrial septal defect that has associated pulmonary vascular changes. Even after cardiac catheterization the only difference that could be noted has been the presence of left axis deviation. Such deviation, although common, is not an invariable finding, however, for we have seen proved instances where it was not present.

[ventr]icular valves are therefore incompetent, and evidence of insuf[fici]ency of the mitral and tricuspid valves results These patients may [hav]e features simulating a large atrial septal defect. Splitting of the [sec]ond sound may be wide and relatively fixed with respiration (Fig [43]1) Pulmonary arterial hypertension is a common sequel, and when [thi]s present patients present the typical features of pulmonary hyper[ten]sion (Figs 432 through 435) On auscultation the second heart [so]und is accentuated often greatly so Splitting of the second heart [so]und is more apt to be close increasing slightly with inspiration [(F]ig 433) as opposed to the fixed, wide splitting of the usual uncom[pl]icated atrial septal defect A systolic murmur generally short, is [h]eard over the pulmonary area In addition a pulmonary ejection [so]und may be present Usually depending on the degree of hyperten[si]on *in the pulmonary circuit a diastolic blowing murmur may be* [h]eard along the left sternal border suggesting insufficiency of the [p]ulmonary valve (Figs 431, 432 434) At the apex, a pansystolic [m]urmur of grade III to IV intensity is common, and a diastolic

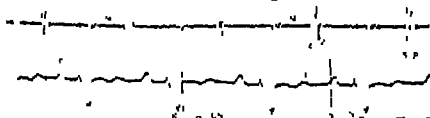


FIG. 43[3] Same patient as Fig 432 Illustrates closely split accentuated second sound which widened slightly with inspiration

rumble is frequently heard suggesting increased flow across the tri[illegible] ... [illegible] liographic ... [illegible] th pulmo[illegible] ... [illegible] Also, as ... [illegible]

[illegible] [congenit]al heart lesion commonly have this type of defect.

ANOMALOUS PULMONARY VENOUS DRAINAGE

The pulmonary veins carrying oxygenated blood from the lungs normally empty into the left atrium Congenital anomalous pulmonary venous drainage may occur so that the veins empty into the right side of the heart via the superior vena cava or left innominate coronary sinus right atrium portal veins ductus venosus or inferior vena cava. When all of the venous drainage is to the right side it is known as total anomalous pulmonary venous drainage, and when a

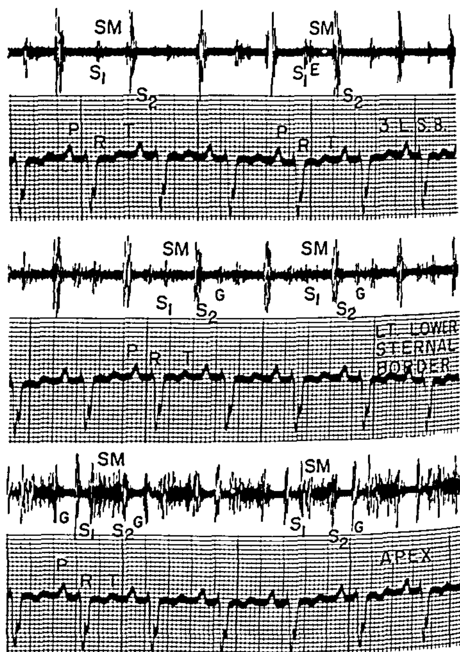


FIG. 432. A 25 year old man with atrioventricular canal associated with pulmonary hypertension. Left pulmonary artery pressure 120/58 mm. Hg. Second [illegible]

[illegible] mation gallop of atrial and ventricular gallops. Had subacute bacterial endocarditis; successfully treated.

the upper ventricular septum due to partial or complete failure of endocardial cushions to fuse during fetal life. This represents a continuous persistent defect involving portions of the lower part of the atrial septum and upper part of the ventricle. The cusps of the atrio-

entricular valves are *therefore incompetent*, and evidence of insuf- … mitral and tricuspid valves results These patients may … t. Splitting of the … respiration (Fig 431) Pulmonary arterial hypertension is a … sequel, and when it is present patients present the typical features of pulmonary hyper- … (Figs. 432 through 435) On auscultation the second heart … is accentuated often greatly so Splitting of the second heart sound is more apt to be close, increasing slightly with inspiration (Fig 433) as opposed to the fixed wide splitting of the usual uncomplicated atrial septal defect A systolic murmur, *generally short*, is … over the pulmonary area In addition a pulmonary ejection sound may be present Usually depending on the degree of hyperten sion in the pulmonary circuit a diastolic blowing murmur may be heard along the left sternal border suggesting insufficiency of the pulmonary valve (Figs 431 432 434) At the apex a pansystolic … of grade III to IV intensity is common and a diastolic

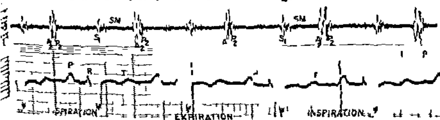


FIG. 433. Same patient as Fig 432 Illustrates closely split accentuated second *sound which widened slightly with inspiration*

rumble is frequently heard suggesting increased flow across the tri cuspid and mitral valves (Figs 431 434) The electrocardiographic findings may be similar to those of atrial septal defect with pulmo nary hypertension showing right ventricular hypertrophy Also as in ostium primum defect left axis deviation may be present as a clue suggesting a different anatomic type of defect from the usual atrial septal defect. It is of interest that Mongoloid children who have a con genital heart lesion commonly have this type of defect.

ANOMALOUS PULMONARY VENOUS DRAINAGE

The pulmonary veins carrying oxygenated blood from the lungs nor mally empty into the left atrium Congenital anomalous pulmonary venous drainage may occur so that the veins empty into the right side of the heart via the superior vena cava or left innominate coronary sinus right atrium portal veins ductus venosus or inferior vena cava. When all of the venous drainage is to the right side it is known as total anomalous pulmonary venous drainage, and when a

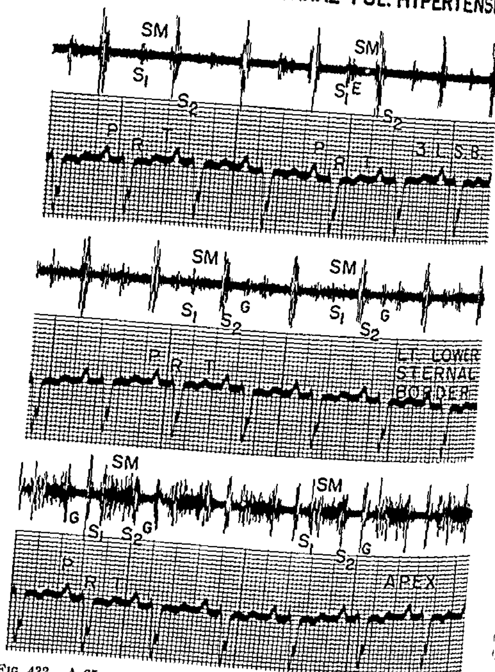
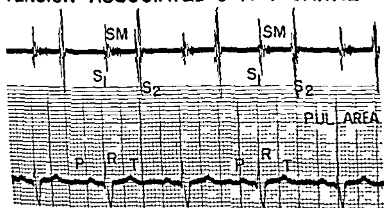


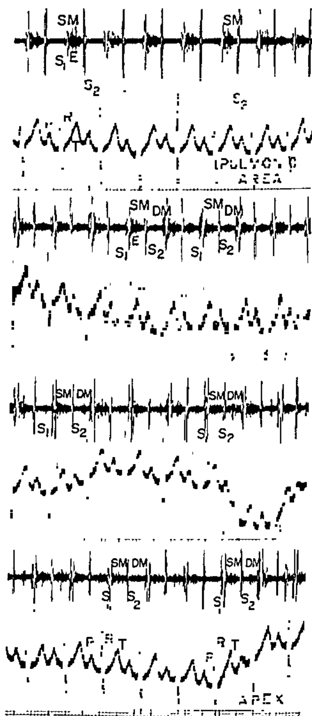
FIG. 432. A 25 year old man with atrioventricular canal associated with pul[illegible] pressure 120/58 mm. Hg. Second [illegible] upper tracing). Note faint systolic murmur (SM) and prominent diastolic gallop [illegible] apex (lower tracing) had pansystolic [illegible] gallop (G) which probably represented summation gallop of atrial and ventricular gallops. Had subacute bacterial endocarditis; successfully treated.

the upper ventricular septum due to partial or complete failure of endocardial cushions to fuse during fetal life. This represents a continuous persistent defect involving portions of the lower part of the atrial septum and upper part of the ventricle. The cusps of the atrio-

ortion is so deviated, it is known as partial anomalous venous drain ge. Symptomatology varies depending on the amount of pulmonary enous blood that is entering the right side of the heart. In effect, the ituation is analogous to that of atrial septal defect, and the symp- oms and findings are very similar in a number of respects If there is otal anomalous drainage cyanosis is variable and may be noticeable nly when congestive heart failure is present, or as a terminal event. Most patients with the total type of drainage die during the first years of life but a few have been reported who reached adulthood There fore most adults will have partial pulmonary venous drainage, which will, as stated, simulate atrial septal defect. In fact a significant number of these patients also have an associated atrial septal defect (Figs. 438, 439, 440) or patent foramen ovale The auscultatory find

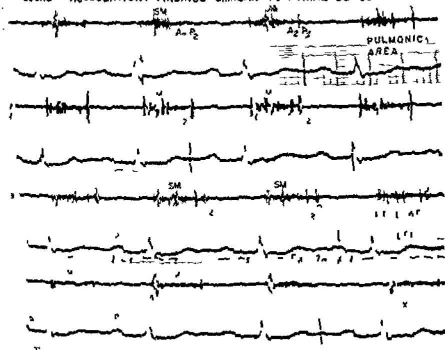


s likewise are similar to those of atrial defect a systolic murmur sually grade III) is heard over the pulmonary area (Figs 436 rough 440) The murmur starts in early systole and as a rule does t continue throughout systole but ends after approximately two rds of it. The second heart sound over the pulmonary area is often dely split and the degree of splitting may be fixed with inspiration igs 436 through 439) Occasionally a diastolic rumble is evident at e apex (Fig. 438) The electrocardiograms are like those in atrial fect, with right ventricular conduction delay in the precordial leads ight bundle branch block complete or incomplete) Likewise pul onary arterial vascular changes may occur with resultant pulmo- ary hypertension and right ventricular hypertrophy In such a case e electrocardiogram would show right ventricular hypertrophy The oentgenogram may show some cardiomegaly and in particular an



with diagnosis of atrioventricular canal and pul- catheterization). Over pulmonic area the second ...tuated, closely split. A grade III systolic murmur (SM) was heard ... pulmonic area and third left sternal space. An early ejection sound (E) was evident. Both systolic (SM) and diastolic (DM) murmurs were heard along the fourth left sternal space and apex (third and fourth tracings). (S_2 retouched by artist to show loud sound which photographed poorly)

, to demonstrate the presence of this condition This is illustrated by a 17 year old school girl whose phonocardiogram is shown in Figure 436 Her diagnosis before catheterization was that of atrial septal defect, based on the classic findings of a grade III pulmonic systolic murmur and a wide splitting of the second heart sound which remained fairly fixed with inspiration The pulmonary artery segment was *moderately* enlarged and vascular markings were moderately increased An electrocardiogram showed right ventricular conduction



... *Systolic* murmur (SM) was heard best at third left sternal space but also over other cardiac areas.

delay When cardiac catheterization was done the catheter entered two separate anomalous pulmonary veins In sp to of th ...

... may have minimal anomalous pulmonary venous drainage and present no symptoms or characteristic findings others with significant amounts of anomalous drainage may present a clinical picture similar to that of a large atrial septal defect with the resultant complication of pulmonary vascular changes If cardiac catheterization reveals a tendency toward pulmonary hypertension with in

enlarged pulmonary artery segment. In some patients the cardiac silhouette is altered by the anomalous drainage. If the drainage is in the superior vena cava, the configuration has been described as resembling a figure 8. Pulmonary vascular markings are increased. In

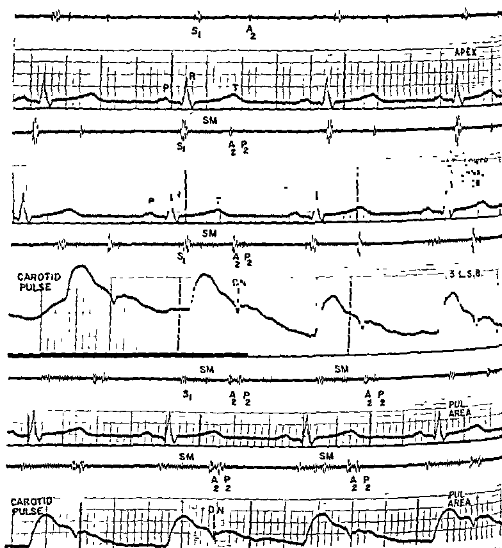
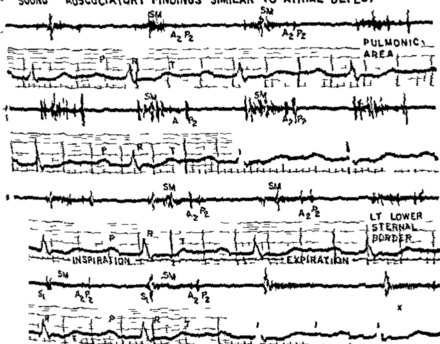


FIG. 436. A 17 year old girl with two anomalous pulmonary veins; demonstrated on cardiac catheterization. The possibility of an associated atrial defect could not be ruled out. Note auscultatory findings similar to those of atrial septal defect. Had f[illegible] o[illegible] th[illegible]

some cases a continuous murmur has been reported, but as a rule the murmur is systolic.

Patients with partial anomalous venous drainage show symptoms so similar to those of atrial defect that cardiac catheterization is

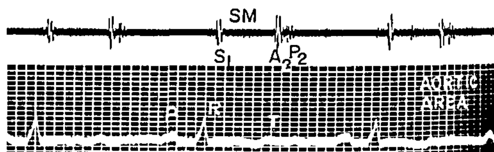
necessary to demonstrate the presence of this condition This is illustrated by a 17 year old school girl whose phonocardiogram is shown in Figure 436 Her diagnosis before catheterization was that of atrial septal defect based on the classic findings of a grade *III pulmonic* systolic murmur and a wide splitting of the second heart sound which remained fairly fixed with inspiration The pulmonary artery segment was moderately enlarged and vascular markings were moderately increased. An electrocardiogram showed right ventricular conduction

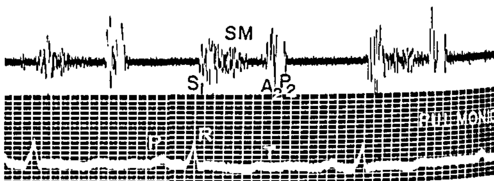


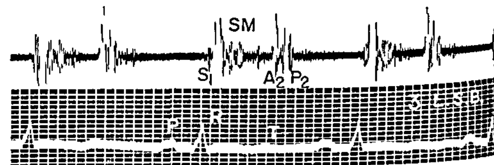
o over other cardiac areas

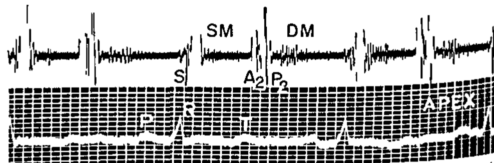
elay When cardiac catheterization was done, the catheter entered wo separate anomalous pulmonary veins In spite of this latter find ng the presence of an associated atrial septal defect or patent fora men ovale could not be ruled out in this case Although it is true that some patients may have minimal anomalous pulmonary venous drain age and present no symptoms or characteristic findings others with significant amounts of anomalous drainage may present a clinical picture similar to that of a large atrial septal defect with the resultant complication of pulmonary vascular changes If cardiac catheteriza tion reveals a tendency toward pulmonary hypertension with in

ANOMALOUS PUL. VENOUS DRAINAGE PLUS ATRIAL SEPTAL DEFECT









reased pulmonary flow, surgery should be considered Reshunting of the anomalous drain nto the left side of the heart has been performed, and with n that surgery in such case roven

Figures 437 and

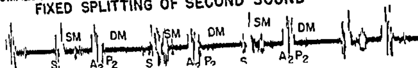
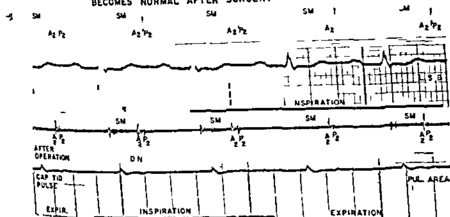




FIG 439 Same patient as Fig 438 showing fixed splitting of second heart sound (A_2P_2) unaffected by respiration. Note systolic (SM) and diastolic (DM) murmurs



persisted after surgery but became fainter

atrial septal defect and partial anomalous venous drainage Surgery was successful in correcting both of her defects A diastolic rumble (a common find ng n ... and/or anomalous pulmonary ... eted as due to ... to the errone ous impression of Lutembacher's syndrome A wide fixed splitting of the second sound became normal after operation (Fig 440)

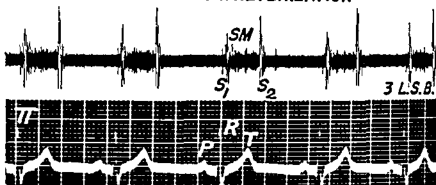


FIG. 441. Man, age 24, with no cardiac symptoms. A grade IV systolic murmur (SM) was heard over the precordium, loudest in the third left interspace.

MURMUR OF INTERVENTRICULAR SEPTAL DEFECT (ROGER'S DISEASE)

SM
S_1 S_2 4 L.S.B.
II
R
P T

SM
S_1 S_2 APEX
II
R
P T

SM
S_1 S_2 PUL. AREA
II
R
P T

FIG. 442. Girl, age 1 [illegible] venous catheterization. E [illegible] at the third and fourth [illegible] cordium (middle and lower tracings).

INTERVENTRICULAR SEPTAL DEFECT

The findings associated with isolated ventricular septal defect ... ral factors the size of the defect, its location and the ... The exact cause of ... The most im- ... ct Most ven ... lar defects are located in the upper membranous portion of the ... The presence of associated congenital defects alters the ... considerably

The Small Uncomplicated Defect (Roger's Disease) These are not uncommon among congenital cardiac abnormalities although the inci- dence is not as high as was thought in the past. With the aid of more

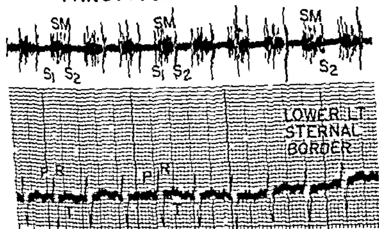


Fig. 442. Baby boy with grade V pansystolic harsh murmur (SM) associated with palpable thrill loudest along the left sternal border at the fourth and fifth interspaces

accurate diagnostic procedures particularly cardiac catheterization, it is apparent that defects such as aortic or pulmonic stenosis, atrial septal defect or mitral insufficiency have been misdiagnosed as ventricular septal defect. A ventricular septal defect of this type is not accompanied by cyanosis or clubbing of the fingers, and is compatible with good health Bacterial endocarditis was a frequent complication in the past accounting for 25 to 40 per cent of the fatalities

On auscultation the significant finding is a moderately loud or very

... est over the pulmonic area (Fig 447) The extent of its transmission

will depend on its intensity, and if it is loud enough it will be heard over the back as well. Obviously, it may closely simulate the murmur of mitral insufficiency or aortic stenosis. In the differential diagnosis the knowledge that a murmur was present shortly after birth or in

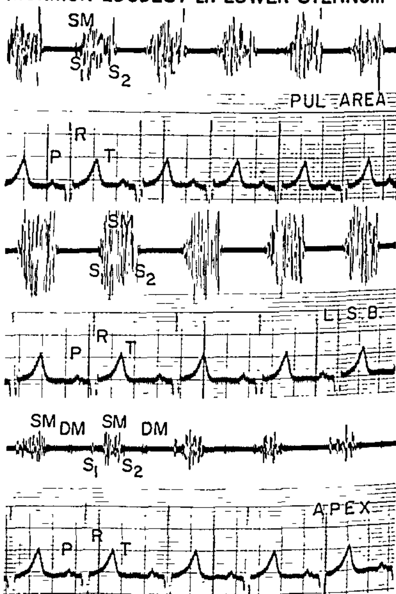


FIG. 444. 7 year old girl with pansystolic murmur (SM) of ventricular septal defect, loudest along lower sternal border (second strip). At apex (bottom tracing) a faint diastolic rumble (DM) also present.

the first few years of life, or that it only appeared after an attack of rheumatic fever, is often helpful in deciding whether the condition is congenital or acquired. It is fair to say that any unexplained moderately loud systolic murmur in the region of the lower precordium,

ecially in the younger patient, should make the physician think of possibility of ventricular septal defect. An absolutely certain diagis can be made by cathete of the heart, as was done in the es referred to above The may show some prominence o cardiogram of a patient with a small, uncomplicated defect is most mmonly normal

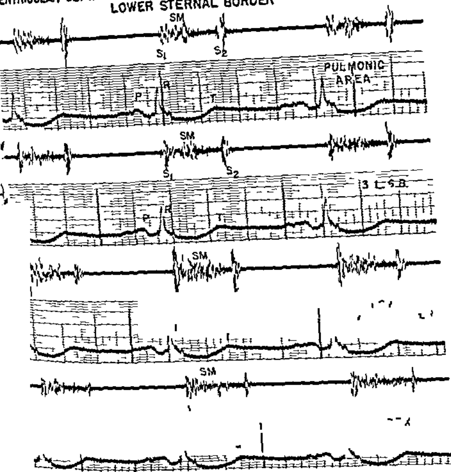


FIG. 445 Man 40 years old with ventricular septal defect Note pansystolic murmur (SM) loudest along left lower sternal border (third strip)

The murmur of ventricular septal defect is rather loud, averaging about grade IV It begins early and generally extends throughout systole (Figs 442 through 448) It is harsh in quality though not to

defect it is otherwise normal or somewhat increased in intensity In

ventricular defect the shunt is from the left ventricle to the right. Following surgical repair, the loud systolic murmur will show a striking decrease in intensity, as shown in Figure 448.

Larger ventricular defects result in greater flow to the right ventricle, pulmonary artery, lungs, and back to the left atrium and left ventricle. This may cause these chambers to enlarge. At the same time, pulmonary resistance may increase, with a resultant increase in pressure on the right side of the heart. If the pressure in the right

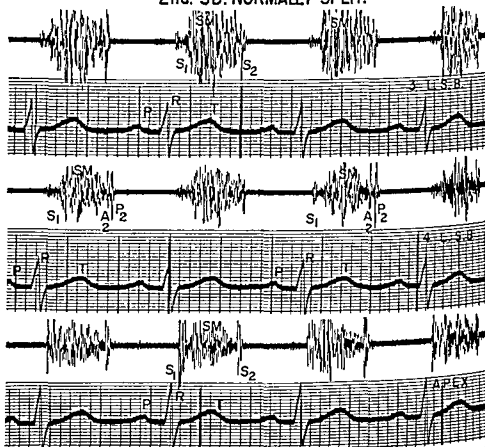


FIG. 446. Two year old girl with pansystolic murmur (SM) of ventricular septal defect, loudest at third left sternal space (upper strip). Second sound (S_2) was normally split, increasing on inspiration (second strip).

ventricle rises sufficiently, approaching or equaling the left ventricular pressure, cyanosis may be evident. In larger defects the systolic murmur likewise varies with these progressive changes. It may be loud with a small defect, and generally, with the larger defects, becomes fainter and may be of shorter duration, ending after midsystole. An early diastolic murmur of insufficiency of the pulmonic valve can result. Also, in some cases, a diastolic rumble is heard on careful auscultation at the apex (Figs. 444, 448, 449), presumably due to increased blood flow. The pulmonary second sound is accentuated

a result of pulmonary hypertension, and is generally more closely
it. With inspiration however, the splitting may widen in normal
hion (Fig 450)

The x ray picture varies with the small defect. There may be no

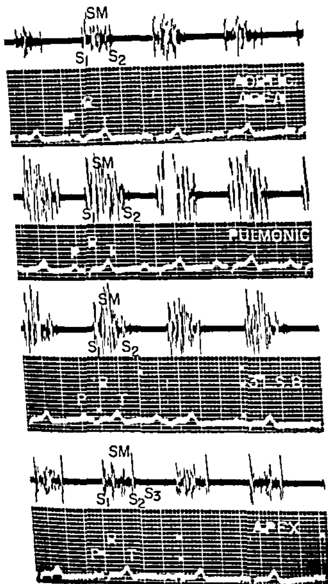
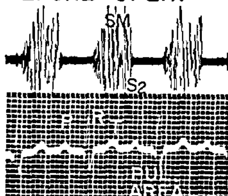
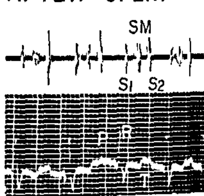


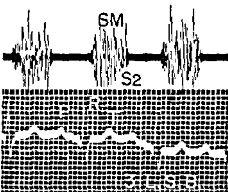
FIG. 447 Girl age 10 subsequently had operation for ventricular septal defect. Had loud pansystolic (SM) murmur loudest over pulmonic area (second strip) and well transmitted over entire precordium. At apex (lower tracing) note third sound (S_3) in addition to first (S_1) and second (S_2) sounds

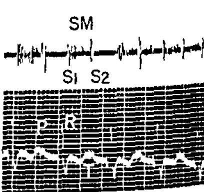
VENTRICULAR SEPTAL DEFECT

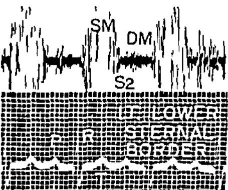
BEFORE OPER. AFTER OPER.











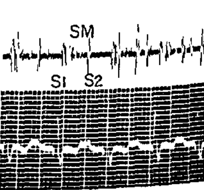


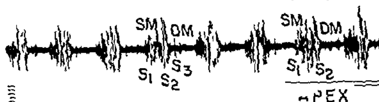
FIG. 448. Boy, age 7, with [illegible] lumn) and [illegible] of murmurs in all areas.

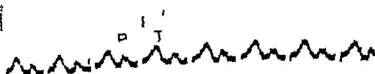
apparent abnormality or only slight pulmonary artery segment enlargement. With larger defects cardiomegaly, associated particularly with enlargement of the left atrium, right ventricle and the pulmonary artery, may be evident. The electrocardiogram will be normal with small, uncomplicated defects; but will show one of a variety of abnormalities with larger defects, including right ventricular hypertrophy, left ventricular hypertrophy, combined right and left ven-

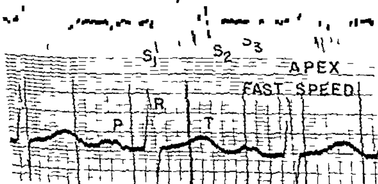
/ENT SEPTAL DEFECT–DEFORMED TRICUSPID VALVE (OPERATION)–DIAST RUMBLE AT APEX











tricular hypertrophy, and conduction defects such as right bund[le] branch block, first degree block, etc.

Ventricular Defects Deforming the Aortic Valve. A ventricul[ar] defect may be so located that it results in a deformity of the aort[ic] valve, producing aortic insufficiency (Fig. 451). This often represent[s] a source of much confusion in diagnosis. Patent ductus arteriosus ha[s] been erroneously diagnosed, and such patients have been subjected t[o] ill-advised operations. Even after cardiac catheterization this confu-sion may exist, as illustrated by the following experience.

A patient was referred to our hospital for operation on patent ductus. A cardiac catheterization performed at the referring hospital revealed findings consistent with patent ductus arteriosus, as evidenced by th[e] "step-up" in oxygen content in the pulmonary artery. The operation re-vealed no ductus, however. Another cardiac catheterization after sur-gery again showed the increase in oxygen in the pulmonary artery. However, a review of the phonocardiogram in this case did not show the typical envelopment of the second heart sound seen with patent

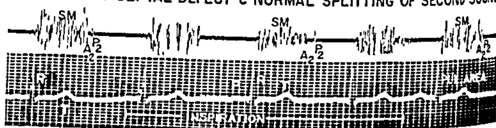


FIG. 450. Same patient as Fig. 447, showing in addition to typical pansystolic murmur (SM) a normal splitting of second sound (A_2P_2) that normally increased with inspiration.

ductus arteriosus. A comparison of the typical murmur of patent ductus arteriosus and the murmur of a septal defect that produces aortic insufficiency is shown in Figure 452. The actual peak in inten-sity was more nearly in midsystole. Since that time we have paid more attention to the phonocardiogram and have found other cases of high interventricular septal defects that simulate patent ductus ar-teriosus. As with the patient described, the peak of the murmur con-sistently was earlier and did not envelop the second sound as occurs in classic patent ductus.

The diagnosis of interventricular septal defect may not be suspected in these cases because of the presenting features of aortic insufficiency resulting from the deformed aortic leaflet. The total cardiovascular evaluation of such patients, however, should suggest the correct diag-nosis. The important features are the x-ray findings of an enlarged pul-monary artery segment, some electrocardiographic evidence of right ventricular hypertrophy (the peak of the murmur being in midsystole rather than enveloping the second heart sound), and an additional point

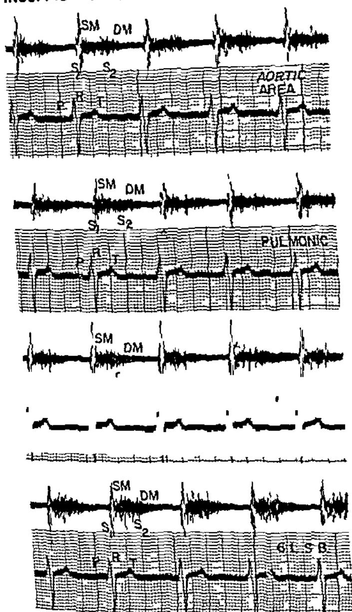
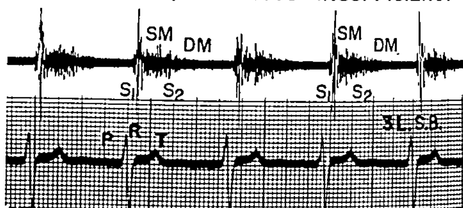


Fig. 451 A 30 year old man with ventricular septal defect deforming aortic

d
d

peak in midsystole and did not envelop second sound.



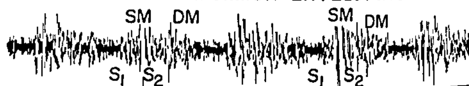
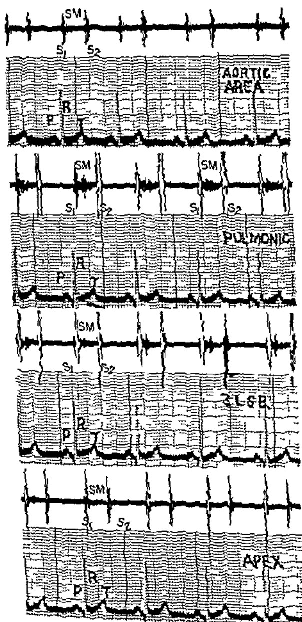


FIG. 452. Same patient as in Fig. 451 (upper tracing) compared with another patient with typical patent ductus murmur (lower tracing). Same patient as Fig 377. Note systolic murmur in top tracing had peak in midsystole, as compared with late systolic component enveloping second sound in lower tracing.

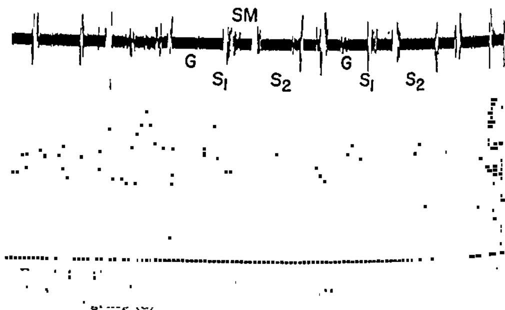
that the murmur is heard better along the third and fourth interspaces, rather than over the pulmonary area as with patent ductus arteriosus.

Ventricular Septal Defect with Pulmonary Hypertension (Eisenmenger Complex)

Pulmonary hypertension and ventricular septal defect are the main features of the so-called Eisenmenger complex. Whether there is an actual overriding of the aorta in such cases is debatable. For all practical purposes, blood can easily be shunted from the right ventricle through the high ventricular defect into the aorta, regardless of whether there is overriding of the aorta. There generally is considerable restriction of physical activities. Moderate cyanosis and some clubbing of the fingers and toes may be present, because there is a right to left shunt. Pulmonary engorgement also is present. The onset


EISENMENGER COMPLEX
SM
S1
S2
AORTIC AREA
P
R
T
PULMONIC
3 L S B
APEX

of cyanosis may be late compared with that of tetralogy of Fallot, and at first may be present only after effort. The x-ray examination shows cardiac enlargement, particularly right-sided, a prominent pulmonary segment, and increased vascularity of the lungs, except when advanced pulmonary hypertension develops. In the latter instance, the periphery of the lung fields may become clear. On physical examination, a right systolic impulse or "lift" may be felt along the left sternal edge, and prominent "a" waves noted in the jugular pulse of the neck. On auscultation a systolic murmur is present that is heard best along the second or third left sternal interspace (Figs. 453, 454). The murmur varies, but is generally of grade III to IV intensity. It is usually short, starting early in systole, and often ends shortly after



midsystole. Diastolic murmurs may also be present. An early, blowing diastolic murmur of pulmonary insufficiency occurs with the higher degrees of hypertension in the pulmonary circuit. This also is heard best along the third to fourth left sternal interspace. At the apex a rumble is sometimes heard in middle to late diastole, and is presumably due to increased blood flow across the mitral valve.

A very common finding is an accentuated second sound over the pulmonary area (Figs. 453 through 456). The degree of accentuation parallels the extent of the pulmonary hypertension. The second sound is closely split, and the splitting may increase slightly with inspiration. The second sound is also frequently palpable. In addition, over the pulmonary area and the left sternal border, an ejection sound may be heard in early systole (Figs. 455, 456). Patients have been observed whose systolic murmurs, caused by ventricular septal defect,

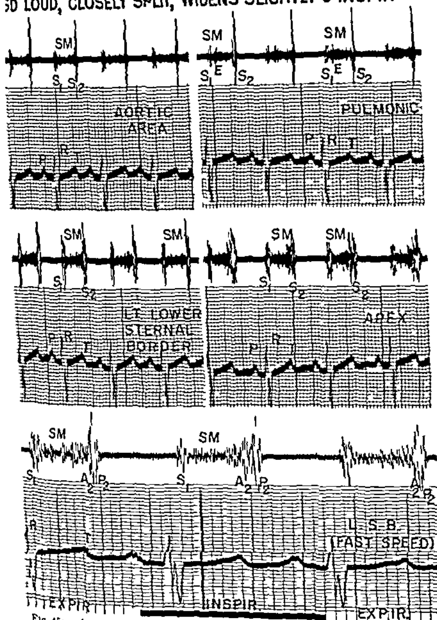


Fig. 45a. A 6 year old girl with ventricular septal defect and pulmonary hypertension no cyanosis or clubbing prominent right ventricular systolic impulse palpable systolic thrill at third and fourth intercostal space palpable second sound

have decreased with the passage of time, owing to the development of significant pulmonary hypertension that increased ventricular pressure. In fact, the murmur from such a defect may become only barely audible, or may even disappear. One can see that, under certain circumstances, only a pulmonary systolic murmur may remain. It is evi-

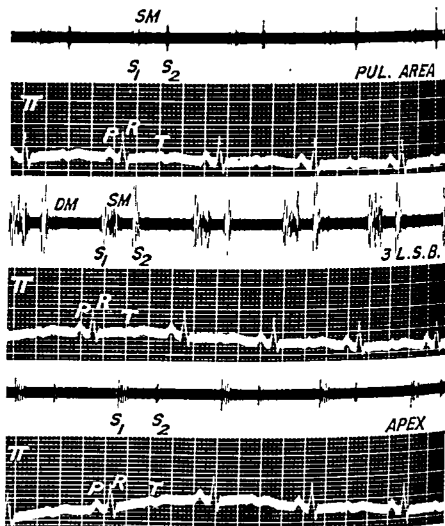


FIG. 456. A 32 year old woman. Findings on cardiac catheterization consistent with Eisenmenger complex. At the third left interspace (middle tracing), a grade III rough, short systolic murmur (SM) and a definite grade II high pitched, blowing diastolic murmur (DM) were heard P_2 accentuated (S_2). Upper and lower tracings taken at pulmonic and apical regions respectively.

dent that the severity of a given patient's defect is not dependent on the loudness of his murmur. In fact, the loudest murmurs are usually associated with uncomplicated defects (Roger's disease). An atrial gallop may be present. With the onset of heart failure, a ventricular diastolic gallop rhythm is often heard.

AORTIC OR SUBAORTIC STENOSIS

When this condition is congenital the signs for the most part are similar to those observed in acquired aortic stenosis, but are found in young individuals If it is known that the murmur was present shortly after birth and that there is no history of rheumatic infection the possibility of congenital aortic or subaortic stenosis must be considered whenever the findings resemble those of acquired aortic stenosis

Often a very loud harsh murmur with greatest intensity in the aortic region is present (Figs 457 through 462) A prominent thrill

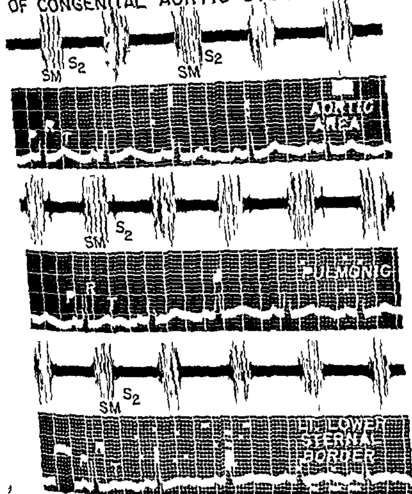


FIG. 457 A 6 year old boy with congenital aortic stenosis proved at postmortem examination. A grade VI harsh aortic systolic murmur (SM) was present which was transmitted widely over the precordium Clinically the second sound (S_2) was louder than recorded. (Because of extreme loudness of murmur volume was reduced during recording)

accompanies the murmur. Although generally loud, the murmur varies in intensity from grade III to grade VI. Sometimes it is heard best along the left sternal border, and, in early infancy, this often leads to its misinterpretation as a murmur of septal defect or pulmonic stenosis. An important differential diagnostic point to keep in mind when the murmur is heard best along the left sternal border is the direction of the palpable thrill. The thrill of pulmonic stenosis radiates

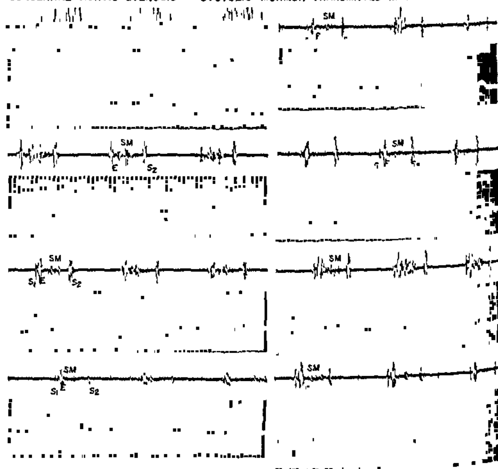


FIG. 458. A 37 year old man with aortic stenosis. Note wide transmission of systolic murmur (SM); loudest over aortic area (left upper tracing) and third right sternal space (second tracing, left column). Note normal or slightly accentuated second sound (S_2) and early systolic ejection sound (E).

toward the left shoulder, whereas that of aortic stenosis radiates toward the right shoulder. The same is true of the murmur. The murmur may be heard at the apex and, if loud enough, over the back also. As a rule, the degree of stenosis is reflected by the loudness of the murmur. No such correlation appears to hold true for acquired aortic stenosis.

The aortic second sound is usually normal, or may even be accentuated (Figs. 458 through 462). Rarely is it absent. Owing to the delayed closure of the aortic valve caused by the stenosis, it may close

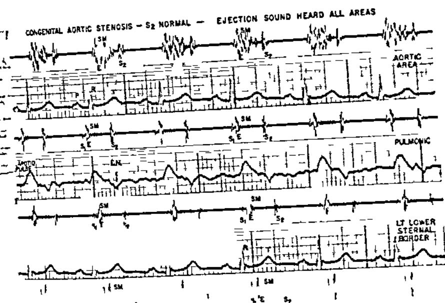


FIG. 459 A 26 year old man with congenital aortic stenosis. Note harsh aortic systolic murmur (SM) heard best over aortic area (upper tracing) Aortic second sound (S_2) was of normal intensity Had prominent ejection sound (E) in early systole in all areas

CONGENITAL AORTIC STENOSIS — NORMAL SPLITTING OF SECOND SOUND

CAROTID PULSE

EXPIRATION

AORTIC

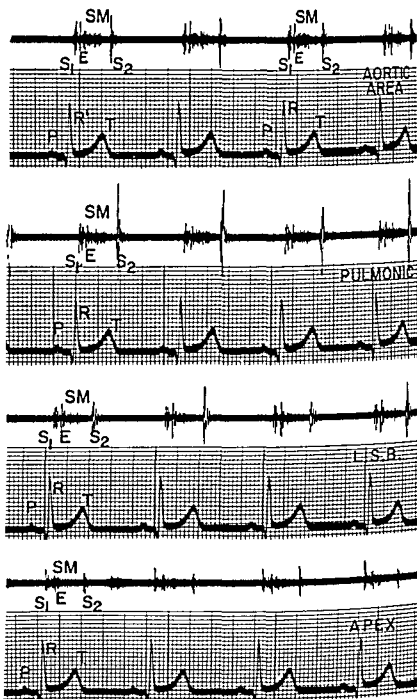


FIG. 461. Man, age 24, with aortic stenosis; probably congenital. In addition to grade IV systolic murmur (SM) heard best over aortic area, an early systolic ejection sound (E) was heard over all areas. Note second sound (S_2) was normal to slightly accentuated.

just before the pulmonic, thus producing a close splitting. If aortic closure coincides with pulmonic, a single second sound results. In each instance, with inspiration, there is a normal widening of the splitting (Fig. 460). If enough delay results from more severe stenosis, aortic

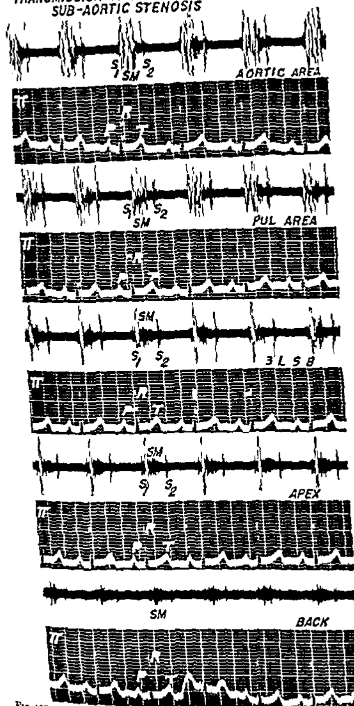


FIG. 462. Boy age 12 with a loud harsh grade VI systolic murmur (SM) loudest in aortic area (first tracing). Murmur (SM) was also transmitted over precordium (second third and fourth tracings) also well heard in back (lowest tracing). Cardiac catheterization showed no evidence of pulmonic stenosis septal defects or left to right shunts. Had prominent early systolic ejection sound at apex.

valve closure occurs after pulmonic closure, and splitting is present. In this latter instance, with inspiration and the coincident delay in pulmonic valve closure, splitting may paradoxically become less, or the sound may become single. Another auscultatory finding is an early systolic ejection sound heard at the apex, over the aortic area and at the carotid artery. An early, blowing aortic diastolic murmur is uncommon.

Although we have found no specific criteria to distinguish aortic valvular stenosis from subvalvular, the diagnosis of congenital aortic stenosis should present no difficulty in the majority of cases. Diag-

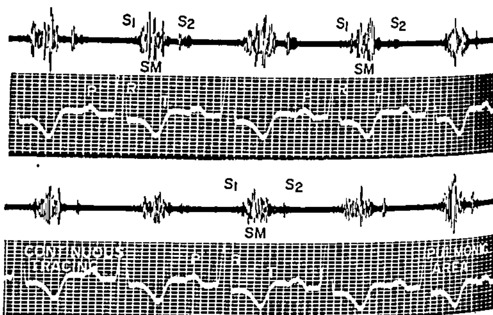


Fig. 463. Boy, age 15, with clinical features suggesting congenital aortic stenosis. At surgery no valvular stenosis was present, but instead a subaortic diaphragm was present and was removed. Note harsh diamond-shaped systolic murmur (SM) and splitting of second sound (S_2).

nosis is nearly certain if there is a comi[illegible] tors: a history of the murmur in infanc[illegible] of a history of rheumatic fever; a loud, [illegible] palpable thrill that is directed rightward; electrocardiographic evidence of left ventricular hypertrophy or of a normal left ventricle; and x-ray evidence of left ventricular hypertrophy without pulmonary artery enlargement. Sudden death can occur in a previously asymptomatic patient.

Subaortic Diaphragm. This rare anomaly, which has all the features of congenital aortic stenosis, was found at operation in a 15 year old boy (Fig. 463). Aortic stenosis had been the preoperative diagnosis, but the aortic valve was normal. Instead a congenital diaphragm

as present below the valve causing obstruction to blood flow in the me manner as aortic stenosis This was corrected by surgery

CONGENITAL AORTIC INSUFFICIENCY

This is very rare as an isolated lesion Even in one of our hospitals here patients with severe aortic insufficiency have been considered possible surgery only two cases out of approximately 600 patients thought to be of congenital origin One of these (Fig 464) was a 7 year old boy who had had aortic insufficiency since birth He was eferred for operation because of progressive signs of cardiac decom ensation The clinical findings in his case were not different from hose of severe aortic insufficiency from other causes There was a hort grade III systolic murmur over the aortic area (Fig 464) The econd heart sound was accentuated over the aortic area, and an early owing diastolic murmur (grade IV intensity) was heard best along e third left sternal interspace At the apex a grade III systolic mur ur was present thought to be due to relative mitral insufficiency, d in addition the transmitted murmur of aortic insufficiency was ident In a well localized spot a faint diastolic rumble (Austin lint) was also present

The only feature in this case that pointed to a congenital origin was he fact that the murmur had been heard since birth A plastic arti icial valve was successfully placed in the first portion of his descend ng aorta He died suddenly approximately five months later, and a postmortem examination was reported to show a congenital separa tion of a portion of the valve ring from its attachment to the aorta

CONGENITAL PULMONIC STENOSIS

Uncomplicated congenital pulmonic stenosis generally presents characteristic features that enable the diagnosis to be made with a high degree of accuracy It is first suspected when a systolic murmur is found at the pulmonary area (Fig 465) This sound is harsh rough in quality and has its maximum peak around midsystole producing a diamond shaped murmur like that of aortic stenosis The sound is gen erally heard best in the second left intercostal space and is trans mitted widely over the precordium It is also heard well in the first left intercostal space and occasionally is loudest over the third left intercostal space If the murmur is over grade III, a palpable thrill is present and is directed toward the left neck and shoulder When the murmur is loud it is heard well at the back but better in the left upper back region than in the right A diastolic murmur of insufficiency of the pulmonary valve is unusual but may occur

The typical loud murmur of pulmonic stenosis usually presents no diagnostic problem Its rough quality simulates the sound of clearing one s throat The fainter murmurs usually signifying a milder degree

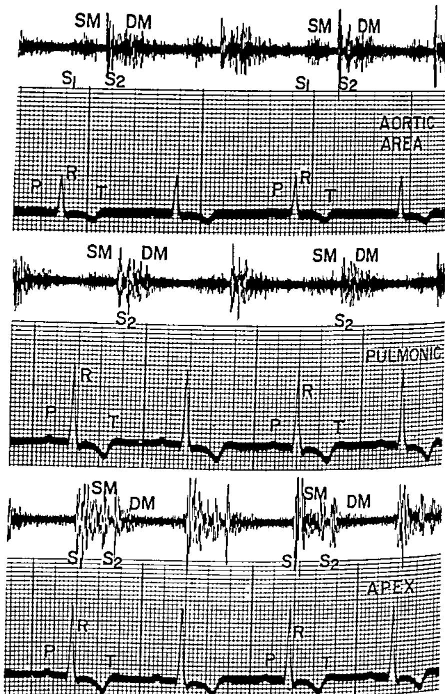
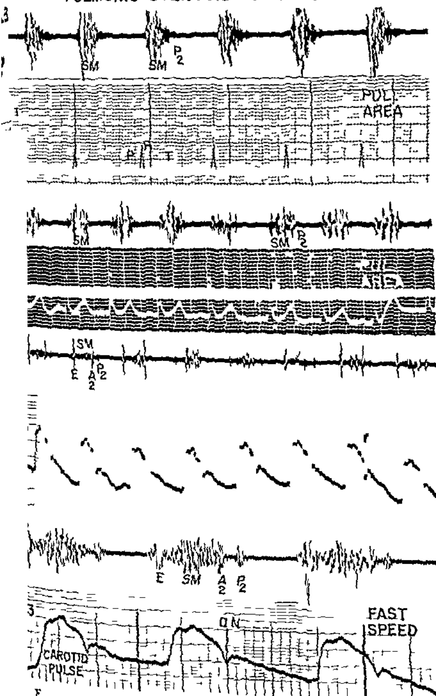


FIG. 464. Boy, age 17, with known aortic insufficiency since infancy. Had classical clinical features of severe aortic insufficiency. Blood pressure 190/0. Had progressive cardiac decompensation; Died suddenly five months after operation for aortic insufficiency. Postmortem reported to have shown congenital separation of the aortic valve ring from its attachment to the aorta. The valve itself was apparently normal.



... e closure (P2) in each

of stenosis, are the ones most difficult to interpret. With moderate or severe degrees of stenosis, the systolic murmur over the pulmonic area usually envelops and obscures the first component of the second sound, the aortic valve closure (Fig. 466). The pulmonic valve closure, because of the stenosis, is delayed and, as a rule, faint. In fact, in a number of these patients one must listen carefully to detect a faint

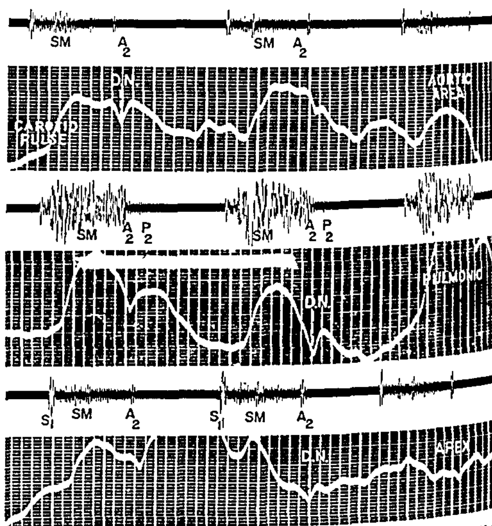
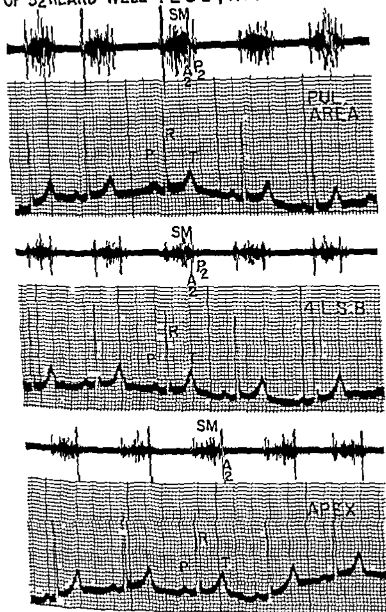


FIG. 466. A 12 year old girl with clinical features of pulmonic stenosis. Note harsh grade V diamond-sh… tends to include aortic co… (P_2) is faint and delayed … and apical areas.

sound well after the ending of the murmur. Actually the second sound is widely split, but the first part, or aortic valve closure, is "covered up" by the prolonged murmur. By listening along the lower left sternal border, the wide splitting may become evident (Fig. 467). In some patients the split widens even further with inspiration (Fig. 468). One might suspect that inspiration would [illegible]ease the degree of



...cond heart sound (A P) was clinically evident because the stenotic murmur was ...ss i ...ense in this area.

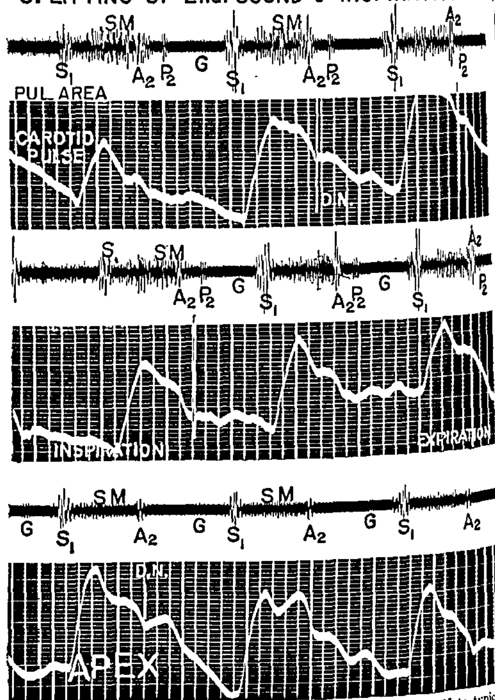
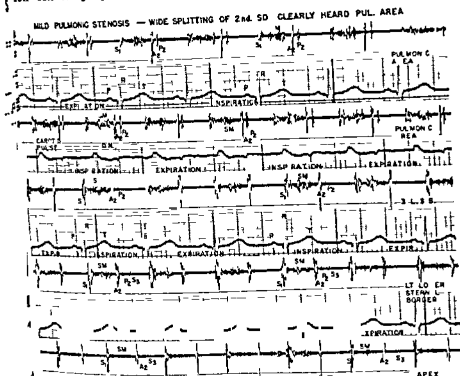


FIG. 468. A 5 year old girl with congenital pulmonic stenosis Note typical systolic murmur (SM) extending to mask the aortic component (A_2) of the second sound (pulmonic area). The pulmonary component of the second sound (P_2) was delayed and faint. Splitting of the second sound (second strip) increased with inspiration and became closer but not single on expiration. Also had diastolic sound (G) in both areas. Child was asymptomatic and had a normal electrocardiogram and x-ray Stenosis was considered to be of mild degree.

splitting if the stenosis were mild and cause little change if it were severe If there is a wide splitting over the pulmonic area, the diagnosis of moderate or severe stenosis is unlikely On the other hand, with milder degrees of pulmonic stenosis, a wide splitting may be evident and thus may simulate atrial septal defect (Fig 469) In such there is usually little change in this wide splitting with inspiration An early systolic sound (ejection sound) is commonly heard



over the pulmonary artery (Fig 465 lower two tracings) In general its degree of loudness tends to correlate with the degree of stenosis Likewise with greater delay in the pulmonary closure sound there is apt to be a greater degree of stenosis

In most patients isolated pulmonic stenosis is valvular in origin Occasionally however the infundibular type as commonly seen in tetralogy of Fallot, has been observed The murmur of valvular pulmonic stenosis is more likely to be heard higher over the pulmonic area than is that of infundibular stenosis The palpable thrill is also

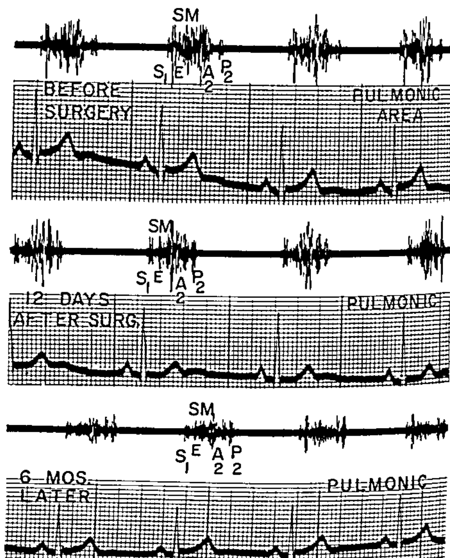


FIG. 470. A 33 year old man. Before surgery (upper strip) had a grade IV harsh systolic murmur with a palpable thrill that radiated toward the left shoulder region. Note early ejection sound (E), harsh, diamond-shaped systolic murmur (SM) masking aortic component of second sound (A_2), and delayed faint pulmonary component (P_2) Twelve days after surgery (middle strip) the auscultatory findings were essentially unchanged except splitting of the second sound was less wide and the pulmonic component (P_2) more easily heard. Six months after surgery (bottom tracing) SM reduced to grade III and splitting of second sound (A_2P_2) had become closer and more easily heard.

higher in valvular stenosis. In fact, both the murmur and the thrill may have their maximum intensity at the third left sternal space in infundibular stenosis.

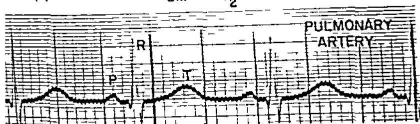
Cyanosis is absent in uncomplicated isolated pulmonic stenosis. If it is present, one should suspect an associated defect, such as atrial or ventricular septal defect. X-ray and fluoroscopic examinations show

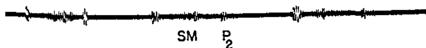
ear lung fields, particularly in the periphery The pulmonary artery egment is often enlarged. The electrocardiogram shows right axis eviation and right ventricular hypertrophy

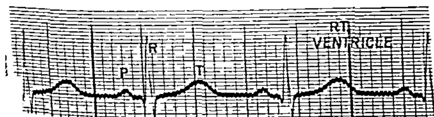
Although the murmur is similar in quality to that of aortic stenosis, ere should be no confusion between the two lesions In pulmonic tenosis the murmur is characteristically over the pulmonic area, and is a palpable thrill that radiates to the left neck or shoulder

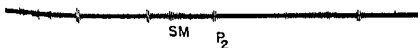
NTRACARDIAC PHONOCARDIOGRAM OF CONGENITAL MILD PULMONIC STENOSIS

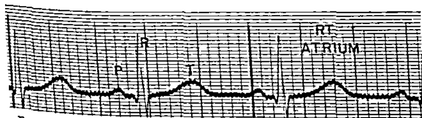












instead of to the right, as with aortic stenosis. The pulmonary artery segment is enlarged in pulmonary stenosis, and not in aortic stenosis. Right ventricular hypertrophy is present in pulmonic stenosis (as evidenced by x-ray examination or the electrocardiogram), compared with left ventricular hypertrophy in aortic stenosis. A left ventricular lift is present with aortic stenosis; a right ventricular lift with pulmonic stenosis. The second heart sound is diminished over the pulmonary area in pulmonic stenosis and is normal or accentuated in con-

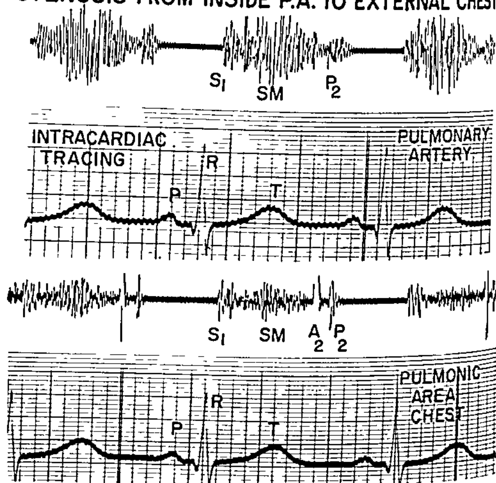


FIG. 472. Same patient as Fig. 471, illustrating decrease in intensity of systolic murmur (SM) of congenital pulmonic stenosis from within the pulmonary artery (upper tracing) as compared with the conventional pulmonary area over the chest wall (lower tracing).

genital aortic stenosis. All of these findings should assist in distinguishing the one lesion from the other.

After surgery for pulmonic stenosis, the systolic murmur usually persists, although decreased in intensity. At times there is little change in the murmur during the immediate postoperative period. With the passage of time, however, the murmur may decrease, as

own in Figure 470 After successful surgery in this patient the pulmonic component of the second sound was less delayed, and the splitting thereby became closer Six months after the operation he continued to show a wider splitting than normal, but the degree was less than before surgery

Intracardiac phonocardiograms demonstrate the precise site of the maximum intensity of the murmur of pulmonic stenosis as shown in Figure 471 The murmur here was loudest in the pulmonary artery just beyond the valve (upper tracing) as compared with sites in the right ventricle and right atrium Figure 472 illustrates the loss of intensity of the murmur from inside the pulmonary artery to the external chest wall Comparable tracings are recorded at the same volume, and it is apparent that a considerable dampening of the murmur takes place by the time one auscults with the stethoscope

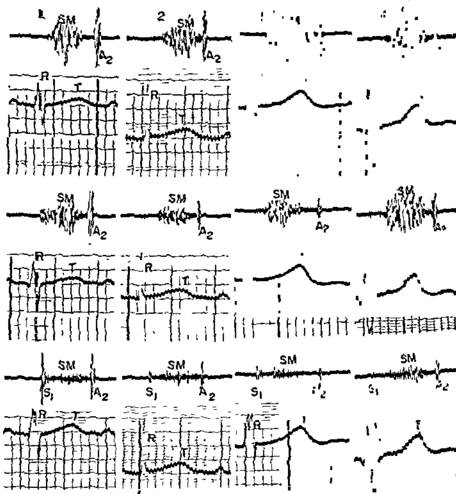
TETRALOGY OF FALLOT

hat the basic defects of this condition are pulmonary stenosis and ... rtrophy largely ... aorta although ... difficult for the ... ally blood from both ventricles may ent... defect. It is also known ... can readily be accomplis... of the aorta. The clinical picture is dependent upon the severity of the two main defects pulmonic stenosis and ventricular septal defect. In adults the most common cause of congenital cardiac cyanosis is tetral-

There generally is considerable limitation in physical activities squatting is common in childhood and congestive failure usually absent. In childhood particularly in infants syncopal episodes may be a presenting feature and death may occur during such an attack Syncopal episodes may also be a prominent complaint in the adult patient (Fig 481, upper tracing) Subacute bacterial endocarditis is uncommon, but brain abscess is not rare Young individuals in particular are prone to respiratory infections against which there is less than the usual resistance One of our patients a young girl with proven tetralogy of Fallot was recently admitted to the Emergency Room with extensive hemoptysis and died shortly afterwards Postmortem examination revealed extensive hemorrhagic pneumonitis

presumably of viral origin. This, incidentally, occurred during an Asian Flu epidemic.

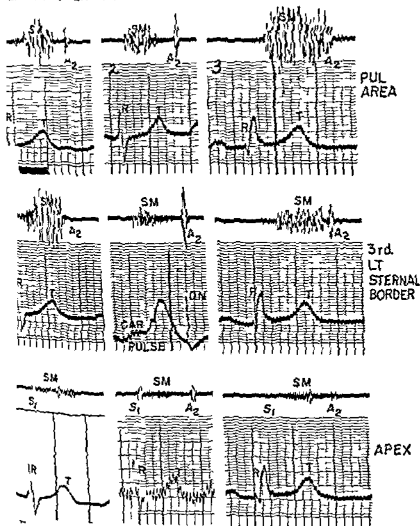
A diagnosis of tetralogy of Fallot must be considered in any patient with marked cyanosis and clubbing of the fingers and episodes of dyspnea—especially if the heart shows any abnormality. The x-ray will show some enlargement of the right ventricle and no engorge-



sound is single in each case, since the pulmonic valve closure was not heard.

ment of the pulmonary vessels. The peripheral lung fields are avascular, and the main pulmonary artery segment is small. On x-ray examination a typical boot-shaped heart, described as "*coeur en sabot*," is present, and a right aortic arch may be noted in approximately 25 per cent of the cases. A systolic murmur is generally present, and is usually of grade III to IV intensity (Figs. 473–479), although at times it be louder or fainter. As a rule, it is heard best at the third left sternal space, but it may be heard equally well over the pulmonic

[?]a or at the fourth left sternal space The origin of this murmur is [beli]eved to be the pulmonary stenosis and most cases of *tetralogy* have [i]nfundibular type of stenosis A small number have a purely valvu



[?]).

[?]s have combinations of infundibular the *systolic murmur of tetralogy* is an isolated purely valvular stenosis

A distinct single second sound (Figs 473 through 479) is heard over the pulmonary *area and along* the left sternal border, and is caused

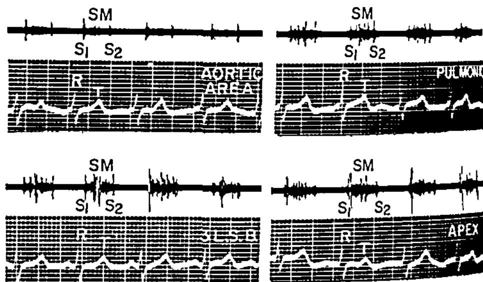


Fig. 475. Boy, age 6, with proven ventricular septal defect plus pulmonic stenosis. Note systolic murmur (SM) and single second sound (S_2). Systolic murmur (SM) heard best at left sternal border (left lower tracing).

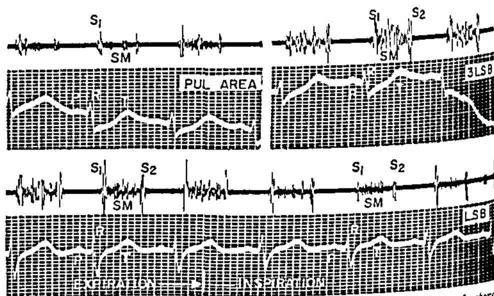


Fig. 476. Boy, age 9, with proven ventricular septal defect and pulmonic steno-
... sternal border (upper right

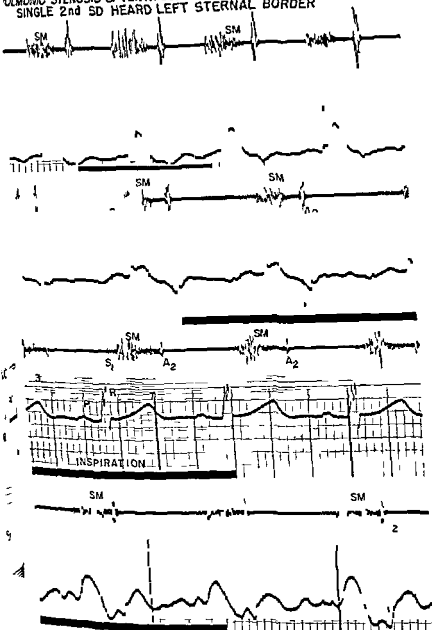


FIG. 477 Composite of four patients with proven pulmonic stenosis and ventric ular septal defect. Systolic murmur (SM) occupied first two-thirds of systole thereby not masking aortic closure of second sound (A₂) Pulmonic valve closure was not h ard therefore second sound was single. With *inspiration no splitting of* second sound noted in any patient.

by aortic valve closure. This second sound is usually well heard, being of moderate or even accentuated intensity. The pulmonic valve closure is rarely heard with the typical type of tetralogy, because its closure is so faint. Therefore, a prominent splitting of the second heart sound would be a point against the diagnosis of uncomplicated tetralogy of

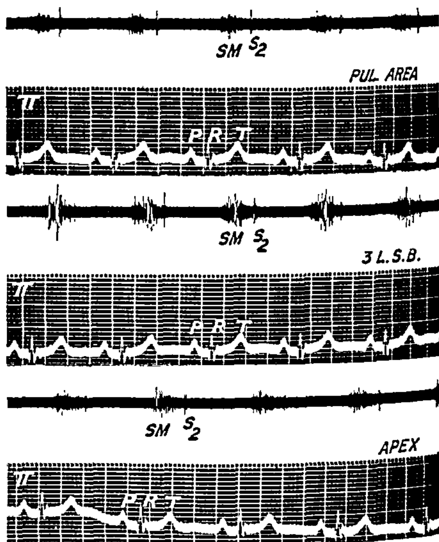
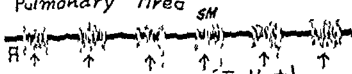
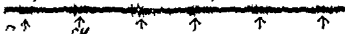


FIG. 478. Woman, age 26. A grade IV systolic murmur (SM) was heard in pulmonic area (upper tracing). Grade V systolic at third left interspace (middle tracing) and grade III systolic at apex (lowest tracing).

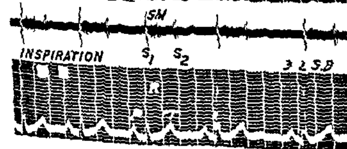
Fallot. A diastolic murmur is not present. The first heart sound at the apex is generally unremarkable, but in some cases an early sound may be heard in systole that is thought to be an ejection sound. This may be misinterpreted as being a splitting of the first heart sound. In the typical tetralogy, the pulmonic stenosis has been of the more severe type, and the septal defect has been large.











ugh systolic mur left interspaces boy with a grade Note decrease in
intensity of heart sounds (S_1 S_2) and murmur (SM) with inspiration

by aortic valve closure. This second sound is usually well heard, being of moderate or even ac[illegible] is rarely heard with t[illegible] is so faint. Therefore, [illegible] would be a point against the diagnosis of uncomplicated tetralogy of

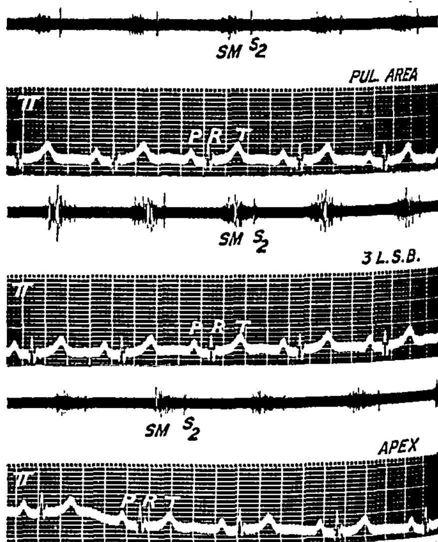
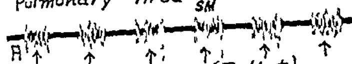
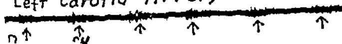


FIG. 478. Woman, age 26. A grade IV systolic murmur (SM) was heard in pulmonic area (upper tracing). Grade V systolic at third left interspace (middle tracing) and grade III systolic at apex (lowest tracing).

Fallot. A diastolic murmur is not present. The first heart sound at the apex is generally unremarkable, but in some cases an early sound may be heard in systole that is thought to be an ejection sound. This may be misinterpreted as being a splitting of the first heart sound. In the typical tetralogy, the pulmonic stenosis has been of the more severe type, and the septal defect has been large.

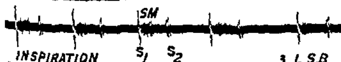
TETRALOGY OF FALLOT — 2 CASES PROVED BY CARDIAC CATHETERIZATION









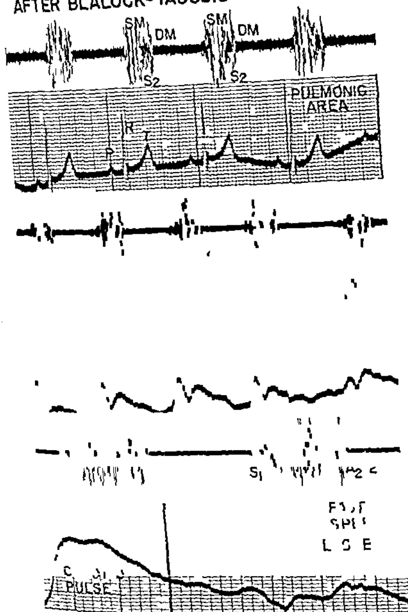


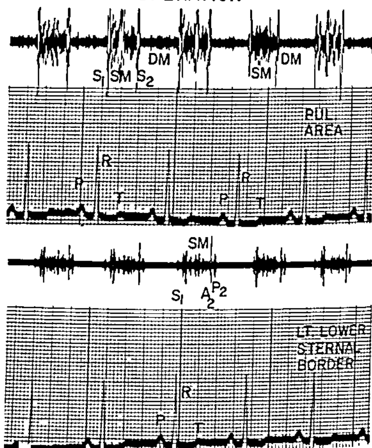


w ic murmur (SM) heard best at the third left interspace Note decrease in ntensity of heart sounds (S_1 S_2) and murmur (SM) with inspiration

The recognition of this condition is very important because it is amenable to surgical treatment. In the past, the defects have not been cured by operation, but the dynamics of the circulation have been improved. The main difficulty is that there is an insufficient amount of blood traversing the pulmonary system to become oxygenated. The first attempt to correct this was an anastomosis of a major artery (the subclavian or innominate) to the pulmonary artery (Blalock Taussig procedure). Later the Potts operation was devised which anastomosed the aorta to the left pulmonary artery. Subsequent direct approaches on the pulmonary stenosis were performed by Brock. More recently, open heart techniques involving the direct repair of both the ventricular defect and the pulmonary stenosis are being performed. This open heart surgery is carried out by means of a pump oxygenator and/or hypothermia. Once these techniques are perfected it is anticipated that the direct correction of the anatomic defects will become the procedure of choice.

Following the anastomotic procedures, patients generally have received considerable benefit. An example of this is shown in Figure 480. This 12 year old boy had had a Blalock-Taussig anastomosis nine years previously. Symptomatically he was much improved, and was essentially well. Prior to the operation he had syncopal episodes and marked cyanosis. Although murmurs were heard in both systole and diastole, they were not of the truly continuous type heard with patent ductus. The murmur was mainly systolic (of grade III to IV intensity) and reached its maximum peak in midsystole. The second sound was of moderate intensity, and following this there was a grade II blowing diastolic murmur that extended throughout diastole but reached its maximum intensity in the early part of diastole. The murmurs were heard best at the third left sternal space. A systolic murmur resembled that heard with uncomplicated pulmonic stenosis. An example of the murmur following the Potts procedure is shown in the bottom tracing of Figure 481. This 6 year old girl had had cyanosis, squatting and syncopal episodes before the operation. Two years later, she had definite symptomatic improvement, a decrease in cyanosis and clubbing, and no longer had to squat. As in the previous patient, a continuous murmur was present, but the murmur was different from that of patent ductus. The murmur was mainly systolic in timing, similar to a pulmonic stenosis murmur, and harsh in quality, reaching its peak in midsystole rather than enveloping the second heart sound. A faint, early, blowing diastolic component was also heard that extended throughout diastole and reached its maximum just after the second heart sound. As another example, Figure 481 (upper tracing) shows a 25 year old man with tetralogy of Fallot several years after a Potts anastomosis had been performed. He also had had a history of cyanosis, clubbing and squatting in childhood, and his presenting complaints prior to surgery were those of exertional dyspnea and syncopal episodes. Cyanosis was moderate. His


TETRALOGY OF FALLOT—NINE YEARS
AFTER BLALOCK-TAUSSIG OPERATION
SM
DM
S2
PULMONIC
AREA
R
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P
S1
PULSE



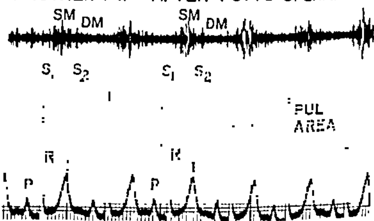
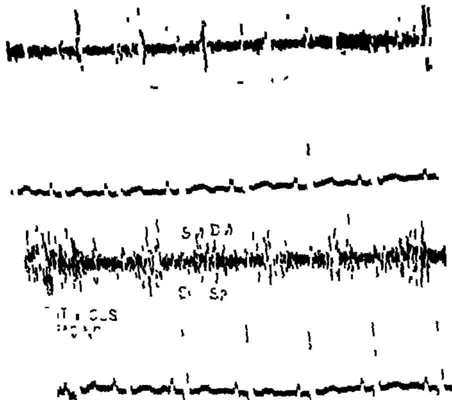


FIG. 481. Composite of two patients with tetralogy of Fallot who had previously ... Potts operation ... old man. Loud systolic mur-

... pulmonary valv...

Lower tracin...
systole and fain...
neither patient ...

murmur was just like the ones already shown following an anastomotic procedure mainly systolic with a maximum peak in midsystole Again a faint diastolic component extended throughout diastole This patient showed signs of cardiac decompensation immediately following surgery and for the first several postoperative months required digitalization and salt restriction. Since that time however,

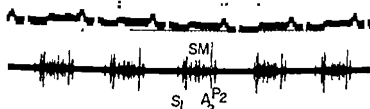


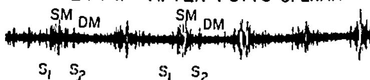
nosis made on cardiac catheterization and angiocardiography

gestive failure has been no problem, and he is working full time a bellhop in a large hotel

Blount and associates Mattingly and others have stressed the extreme variation in this so-called tetralogy of Fallot Grouping of patients with tetralogy of Fallot on the basis of the variations in the two basic anatomic defects (pulmonary stenosis and interventricular septal defect) have been proposed and we feel that this is a logical







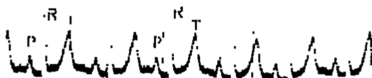


FIG. 481. Composite of two patients with tetralogy of Fallot who had previous
had a Potts operation. Upper two tracings ... A 25 year old man. Loud systolic m

pulmonary valve
Lower tracing
systole and fain
neither patient

Exercise tolerance is better in this group Operative correction should be aimed primarily at closure of the ventricular septal defect, for relief of the pulmonic stenosis alone would probably result in little nefit. Again with the newer techniques it is anticipated that this group likewise will receive great benefit from surgery

PULMONIC STENOSIS + VENTRICULAR SEPTAL DEFECT

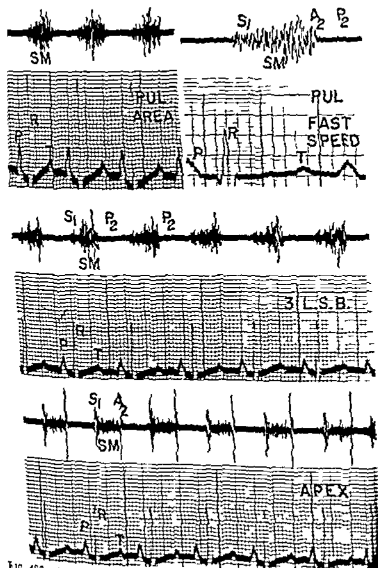


FIG. 455 —

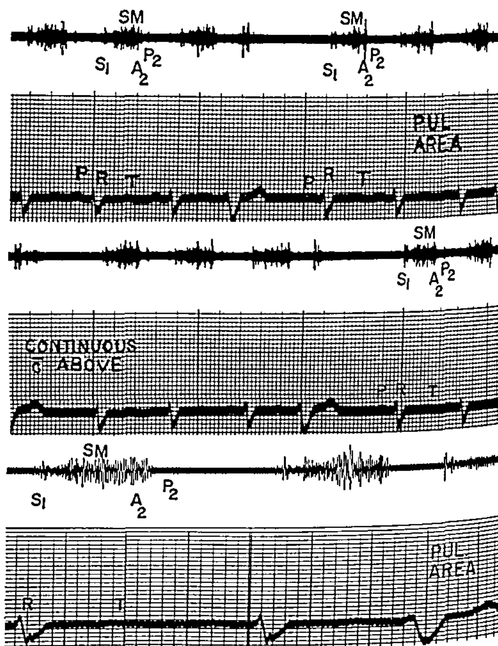
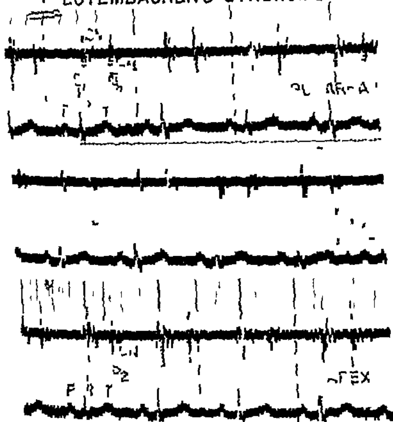


FIG. 485. A 39 year old mother of three, with previous cured subacute bacterial endocarditis. Also had syncopal ep (SM) with palpable thrill radiated sound not heard over pulmonic are: second sound (P_2). Along third or fourth left sternal space, not shown on pho... cardiogram, a short grade II early, blowing diastolic murmur heard.

septal defect and a milder pulmonic stenosis (Fig. 483). Such patients are generally not cyanotic, but have larger hearts with more vascular lung fields than the patient with "classical" tetralogy. The electrocardiogram in such cases would be consistent with that of ventricular septal defect, with right, left or combined ventricular hypertrophy.

A case recently seen had findings suggestive of and compatible with th interatrial septal defect and rheumatic heart disease Cardiac nlargement with *left* and *right* atrial enlargement and marked dila tion of the pulmonary artery and its branches were present Right entricular hypertrophy was evident in the electrocardiogram A



ect proved on cardiac catheterization
matic fever Over pulmonic area and
addition to a third sound or possible
er The possibility of Lutembacher's
possible

ld systolic murmur grade III to IV was heard at the apex and creased on *inspiration* A so
of an opening snap was
rumble A systolic ejec
and the second sound
ntly accentuated and split but *not as* widely as the *splitting* mmonly present with atrial defect alone *Along the left sternal bor* er a grade III early blowing diastolic murmur was present Calcium

A final group would be those patients with severe pulmonic stenosis and a small ventricular septal defect. The presenting symptoms and findings would be those of pulmonic stenosis (Figs. 484 through 486). Surgery in such cases would be mainly directed at the correction of pulmonic stenosis.

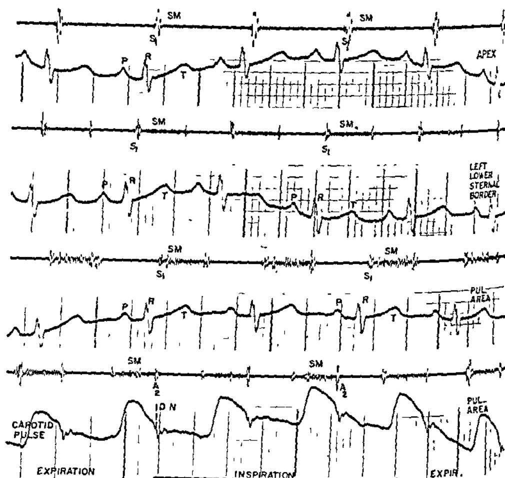


FIG. 487. 11 year old girl with proven pulmonic stenosis and ventricular septal defect No cyanosis. Had systolic murmur (SM) over pulmonic area (lower two strips)

LUTEMBACHER'S SYNDROME

This syndrome is less common than was formerly supposed. At one time, the misinterpretation of the apical diastolic rumble which may be present with uncomplicated atrial defects often led to a mistaken diagnosis of organic mitral stenosis. More recent diagnostic studies and cardiac surgery have shown the syndrome to be uncommon. When it is present, the mitral stenosis is probably on a rheumatic basis and is superimposed on a congenital atrial septal defect.

was seen in the mitral valve on fluoroscopy, signifying stenosis, and the patient's history was compatible with rheumatic heart disease (sore throats migratory joint pains) The patient also had an inter-atrial septal defect (proved by cardiac catheterization) and had superimposed valvular mitral stenosis and mitral insufficiency The clues in this case of combined lesions were atypical features of both difficult to explain on one etiology

...ardiogram of a young soldier with an

an opening snap was also present and the second sou...

...lt. Clinically, the latter seemed evident and the possibility of a ...agnosis of Lutembacher's syndrome arose Left heart catheteriza...on studies were not performed, however Another instance of a pa...ent thought to have Lutembacher's syndrome is shown in Figure ...89 Features of both mitral stenosis and atrial septal defect were ...resent.

AORTIC SEPTAL DEFECT (AORTIC-PULMONARY WINDOW)

This is a rare congenital anomaly but it is important in that it simulates patent ductus arteriosus The defect is an opening between the pulmonary artery and aorta just above the aortic valves The murmur is variable It may be of a continuous machinery type and is heard over the pulmonary area (Fig 490) In general it is more likely to be heard a bit lower than the usual patent ductus murmur although this is not invariably so In a case shown in Figure 491 the murmur was continuous in a well localized spot and was particularly well heard in the first left intercostal space Along the lower sternal border and apex a systolic murmur was present, and over the mitral area a diastolic rumble likewise was heard A systolic ejection sound was noted at the apex and also over the pulmonic area This ejection sound was prominent in the first portion of systole The aortic component of the second heart sound was moderately accentuated and the pulmonic component was not accentuated Following an operation (Fig 491) the diastolic component of the continuous murmur disappeared as did the apical rumble Presumably this rumble was due to increased flow across the mitral valve The systolic ejection sound however was still present It is of interest that, although the murmur was continuous in this case it was not characteristic of classical ductus murmur for the systolic element reached a peak in midsystole rather than in late systole and did not envelop the second heart sound. The diastolic component was much fainter and occurred in the first portion of diastole immediately after the second heart sound

With an increase in the pulmonary vascular resistance and pressure the continuous murmur may be altered as with patent ductus

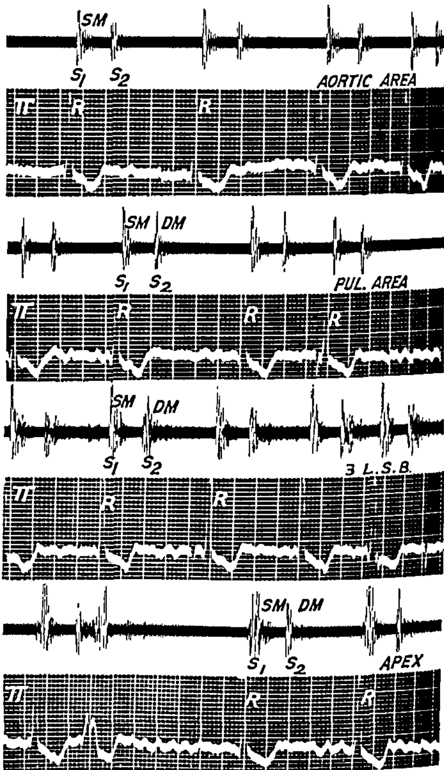
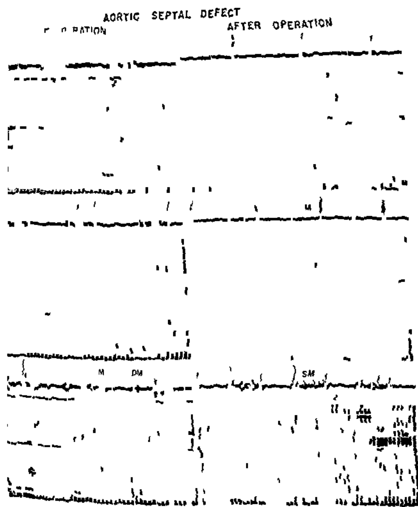


Fig. 489. Man, 28 years old. Had slight systolic murmur (SM) in aortic area (first [illegible]

[illegible]

rumble (DM) without presystolic component. The patient [illegible] rumble was when the rhythm was regular, and a definite crescendo presystolic rumble was present.

machinery murmur would be a point against a diagnosis of far advanced pulmonary vascular changes As with patent ductus, pulmonary hypertension associated with an aortic septal defect should give ...aphic evidence of right ventricular hypertrophy, and



an accentuated, closely split second sound should be heard over the pulmonic area. Some cases are distinguishable only at operation having electrocardiographic x ray cardiac catheterization and physical findings similar to those of patent ductus Although technically difficult, successful surgery has been performed in a few of these rare cases. An example was the case shown in Figure 491

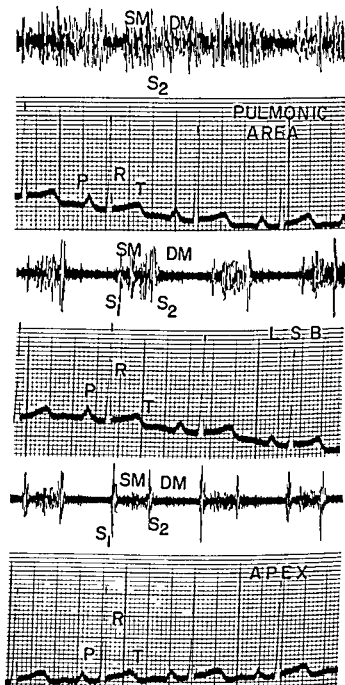
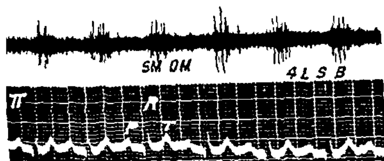


FIG. 490. A 16 year old continuous machinery type was heard best over pulmc... ... (upper strip)... showed left ventricular hypertrophy.

It may eventually disappear, and only a systolic component will then be evident (Fig. 492). Patients may therefore be seen whose systolic murmurs are heard best along the third left intercostal space. The pulmonic component of the second sound is accentuated in such a case. One would surmise that, in older patients, the finding of a continuous

CONGENITAL CORONARY APTERIOVENOUS FISTULA

gical exploration, especially if the murmur is heard best in the pul monary area In some instances, the continuous murmur may be heard in other positions over the precordium, making the diagnosis

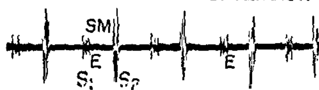


G. 493. A 17 year old boy with a very loud grade VI continuous machinery ... loudest in the fourth left interspace A

s difficult In the case shown in Figure 493 the murmur was loud t at the fourth left sternal space but otherwise was typical of tent ductus There was also a prominent palpable thrill and x ray idence suggestive of patent ductus An exploratory operation re ealed only an arteriovenous fistula involving the wall of the left ven icle, which was rather thin at that area If cardiac catheterization ad been carried out the diagnosis of patent ductus could have been ... it have ... Other ... y arte riovenous fistula and heard best along the lower sternal edge are shown in Figures 494 and 495

The murmur may be heard best along the right sternal border (Fig 496 lower tracing) This would weigh against the diagnosis of patent ductus Cases have been reported where the right coronary artery communicated with the right atrium coronary sinus or the pulmo nary artery The left coronary artery likewise may communicate with the coronary sinus right ventricle pulmonary artery or left ven tricle. In Figure 496 (upper two tracings) a continuous machinery

AORTIC-PULMONARY WINDOW
$\bar{c}$ PULMONARY HYPERTENSION





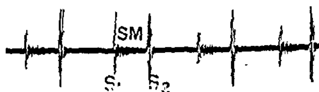


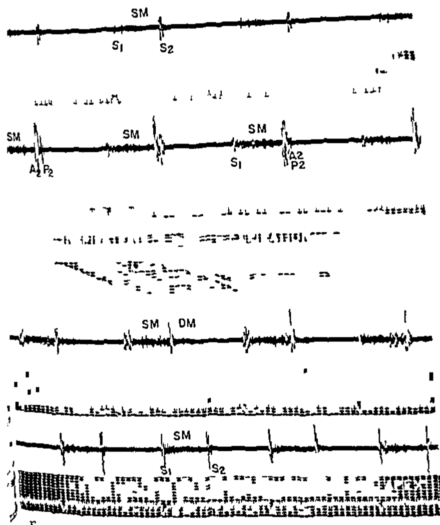






FIG. 492. Gi[illegible] sion diagnosed [illegible] pulmonic and lef[illegible] sound (E) over pulmonic area.

o surgery, patent ductus had been suspected but it was not found when a ductus was sectioned and examined Following surgery the continuous murmur remained as before and the diagnosis postoperatively was uncertain In retrospect (after ruling out other causes of continuous murmur) the most likely diagnosis seemed to be congenital coronary arteriovenous fistula



… er the second sound Systolic murmur at apex (lower strip) and aortic area (upper strip) Note normal splitting of second sound (A_2P_2) coincident with inspiration (second strip)

murmur, grade III intensity, high pitched and heard best with the diaphragm of the stethoscope was present over a well localized spot in the pulmonary area. Cardiac catheterization revealed no evidence of shunt. Surgical exploration was performed, and a coronary arteriovenous fistula was found with the coronary artery entering a venous plexus at the outflow tract of the right ventricle just below the pulmonary valve. The auscultatory feature of this case was atypical in that,

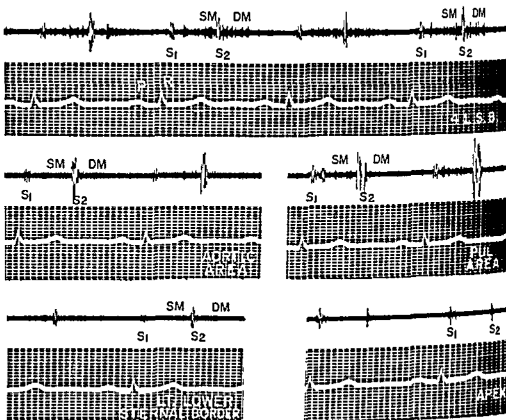


FIG 494. A 40 year old man with coronary arteriovenous fistula. Continuous murmur (SM–DM) with late systolic and early diastolic component noted at fourth left sternal border (upper strip). Murmur was also heard in other areas, including pulmonic (right middle tracing).

although continuous, the murmur started earlier in systole, was of higher frequency and did not "envelop" the second heart sound in the typical patent ductus fashion.

A 25 year old man was noted to have a continuous machinery murmur in a localized spot over his pulmonary area (Fig. 497). His findings were quite similar to the case just described. The murmur was noted to begin early in systole, was of high frequency, heard best using the diaphragm of the stethoscope, and was of grade III intensity. It was poorly transmitted, as in the other patient. Prior

nosis would be more certain It is worthwhile pointing out, however, that even cardiac catheterization findings in those patients where the coronary artery communicates with the pulmonary artery will resemble those of patent ductus It follows that patients will continue to come to surgery with this condition who have *been suspected of having* patent ductus arteriosus

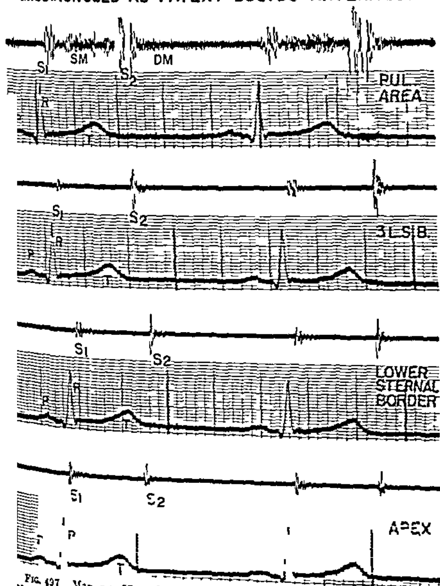


FIG. 497 Man, age 23 with grade III continuous murmur over pulmonic area. Murmur was high frequency type. Systolic component (SM) occupied all of systole diastolic component loudest after second sound (S_2) and continued faintly through out diastole. Patent ductus suspected *prior to surgery* was not present.

The diagnosis of congenital coronary arteriovenous fistula will remain difficult, particularly if the murmur is heard best over the pulmonic area. If the murmur is heard over an atypical area, such as the lower left sternal border or along the right sternal border, the

CONGENITAL CORONARY ARTERIOVENOUS FISTULA
CONTINUOUS MURMUR LOUDEST PUL. AREA

TRAUMATIC CORONARY ARTERIOVENOUS FISTULA



Fig. 496. Composite of two patients with ateriovenous fistula. Upper two strips: Continuous murmur of 37 year old woman Had high pitched, continuous murmur (SM–DM) heard best over pulmonic area. The systolic component occupied all of systole; the diastolic all of diastole. Cardiac catheterization did not reveal patent ductus. At surgery a coronary arteriovenous fistula was found that emptied into the right ventricle just below the pulmonary valve Lower tracing: A soldier, aged 22, with traumatic coronary arteriovenous fistula. Continuous murmur was loudest at right lower sternal border.

confusion with patent ductus arteriosus should not be great. A murmur of high frequency, on the fainter side, and "closer to the ear" by auscultation should at least raise one's suspicion of the presence of this abnormality (Figs. 496, 497). If cardiac catheterization does not reveal any increase in oxygen in the pulmonary artery, the diag-

area), rapid signs of cardiac decompensation, and a lack of enlargement of the pulmonary artery—all of these points would militate against a diagnosis of patent ductus Also, the continuous murmur may not envelop the second sound as it does in a classic case of

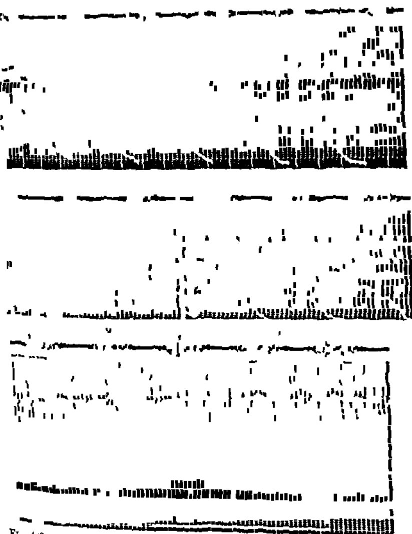


Fig. 498. Man age 28 with ruptured sinus of Valsalva into right ventricle. Continuous machinery murmur (SM-DM) was heard best along left sternal border from pulmonic area to lower middle sternum.

patent ductus Instead it will have a maximum peak of intensity in midsystole and will be heard better in some cases along the right sternal border than at the left (Figs 498 through 502)

Successful operations have been performed on these patients both

CONGENITAL SINUS OF VALSALVA ANEURYSM (WITH AND WITHOUT RUPTURE)

An aneurysm of the sinus of Valsalva may be acquired or congenital. In the past, it was thought that most were acquired and were of syphilitic origin. However, of the six cases we have seen during the past several years, none had had syphilis; all were presumed to be of congenital origin, or a result of an inherited defect of aortic medionecrosis associated with a Marfan's syndrome or a variant of it. As already discussed under Aortic Insufficiency, our attention was first called to sinus of Valsalva aneurysm as a cause of severe aortic insufficiency by observing a patient whose diastolic murmur was heard best along the right sternal border (third and fourth intercostal spaces) and not the left. Further investigation, including thoracic aortograms, revealed the aneurysm in this patient. There was a rightward displacement of the aortic root in this instance. Since then, other examples have been noted; in several the first clue was the "right-sided" murmur of aortic insufficiency.

The diagnosis of sinus of Valsalva aneurysm will probably not be suspected on auscultation unless the deformity is sufficient to produce insufficiency of the aortic valve itself, or unless an actual rupture occurs. An aneurysm of a sinus of Valsalva producing aortic insufficiency generally is accompanied by a diastolic murmur that is heard best along the left sternal border. It is only when "right-sided" murmurs of aortic insufficiency are present that aneurysm of the sinus of Valsalva can be suspected as one of several possible causes. It is obvious that some cases will be called rheumatic aortic insufficiency and not be recognized except at autopsy or after an acute rupture.

A perforation or rupture of the sinus of Valsalva is generally into the right side of the heart (right atrium or right ventricle). The right aortic sinus is most frequently involved; the noncoronary is the next most likely to have aneurysm with rupture. The left aortic sinus is the least likely site. Although a rupture into the right side of the heart is the most common, a perforation into the left side or the pulmonary artery, or even a dissection down into the septum, causing complete heart block, may occur. When rupture takes place, or if a congenital communication already exists, a continuous machinery murmur, or a sound that simulates a continuous murmur (one that has systolic and diastolic components of a "to-and-fro" quality), suggests the diagnosis (Figs. 498 through 502).

When a rupture occurs, the patient is often aware of "something happening in the chest," and he may even note the onset of a noise in his chest. The continuous murmur may be heard best along the left sternal border, and it will simulate the murmur of patent ductus. An evaluation of the total clinical picture, however, should be sufficient to establish the proper diagnosis. The sudden occurrence of a "spell," the appearance of a loud, continuous murmur (except over the pulmonic


CONGENITAL SINUS OF VALSALVA
RUPTURED INTO RT VENTRICLE
S2
AORTIC
AREA
R
P

with and without open heart techniques. The patient whose case is illustrated in Figure 499 had a plastic prosthetic "golf tee" inserted, and the continuous murmur reportedly disappeared after surgery.

Figures 501 and 502 illustrate an instance of proven congenital aneurysm of the sinus of Valsalva with a congenital communication

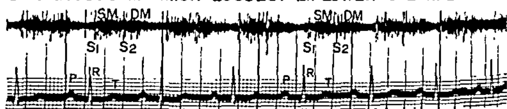


FIG. 499. A 27 year old man who had a sudden occurrence of a continuous machinery murmur (SM–DM) that was heard best at the fourth left sternal space. It was accompanied by a thrill. The diagnosis of a ruptured congenital sinus of Valsalva aneurysm was proved by left heart angiograms and by surgery. The murmur resembled that of patent ductus, but had a systolic component that was loudest in midsystole and that had an atypical location. The continuous murmur was reported to have disappeared after surgery.

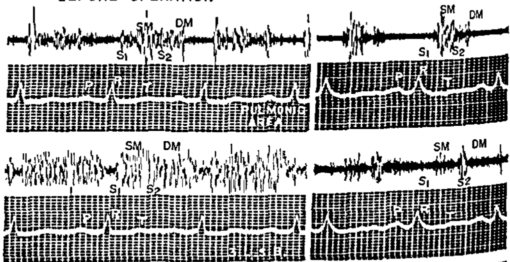


FIG. 500. A 54 year old woman with ruptured sinus of Valsalva. Before surgery had continuous murmur (SM–DM) heard best at third left sternal border (lower left strip). Following operation (right column) using plastic prosthesis, note decrease in intensity of murmur.

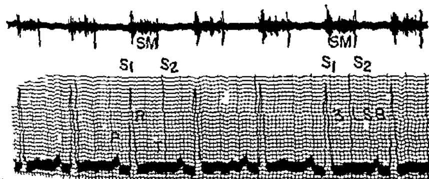
into the right ventricle. This six year old girl had a grade IV to V murmur that was heard best at the fourth left sternal space, although the sound was easily heard over the entire precordium. The murmur had somewhat of a "to-and-fro" quality, although it was continuous, machinery-like and simulated the murmur of patent ductus at the fourth left sternal interspace. The phonocardiograph, however, showed that the peak of the systolic component was closer to mid-

axis deviation or left ventricular hypertrophy, and both are important features Enlargement of the P waves is common

Most of these patients die during the first year of life, although a few survive longer (Figs 503, 504) According to the classification of Edwards they may be divided into two main groups those with transposition of the great vessels and those without transposition.

Tricuspid Atresia with Transposition of the Great Vessels Patients in this category are further subdivided into two types (1) transposition with pulmonary stenosis and a ventricular septal defect (2) transposition with ventricular septal defect but without pulmonary stenosis

Figure 503 illustrates the case of a patient with tricuspid atresia,



transposition pulmonic stenosis and a ventricular defect He was 23 years old and had had deep cyanosis since birth His clinical picture was that of severe cyanosis extreme clubbing of the fingers and toes and decreased exercise tolerance A grade IV systolic murmur was present and was loudest over the third left sternal space It was harsh stenotic in character and was well transmitted over the precordium and over the right carotid artery in the neck A palpable systolic thrill was present which radiated toward the right shoulder A diagnosis of truncus arteriosus was ruled out because the second sound was definitely split, and the patient appeared to belong within the Edwards group having tricuspid atresia with transposition pulmonic stenosis and ventricular septal defect The direction of the palpable thrill was most helpful in establishing the diagnosis as it suggested a stenotic area which if transposed should be pulmonic stenosis The diagnosis

systole, even in this area. The patient had peripheral signs of aortic insufficiency and left ventricular hypertrophy, and these conditions were also apparent fluoroscopically. Patent ductus could be quickly excluded from the differential diagnosis by the clinical features discussed above and, in particular, by the location and character of the murmur. Right heart catheterization revealed an increase in oxygen content in the right ventricle. Sinus of Valsalva aneurysm with a con-

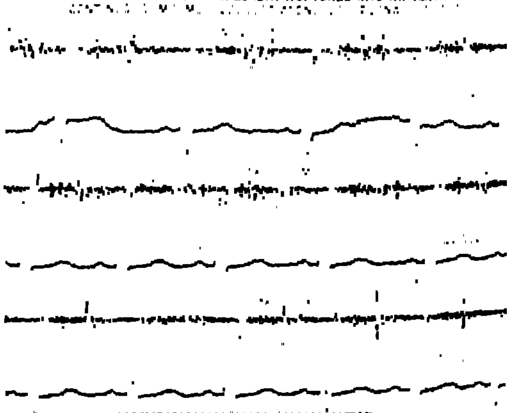


FIG. 502 Same patient as Fig. 501 Tracings taken at faster speed to illustrate configuration of continuous type of murmur (SM DM) loudest along lower left

genital communication into the right ventricle was thus suspected, and the diagnosis was subsequently verified by special angiograms.

TRICUSPID ATRESIA

In tricuspid atresia, which is rare, blood cannot pass directly from the right atrium into the right ventricle. Of necessity, it is shunted through an atrial septal defect or patent foramen ovale. Cyanosis is a prominent feature of this condition and is usually marked. These patients also commonly have a decreased tolerance to exercise, syncope and frequent upper respiratory infections. Clubbing of the fingers and toes is present. The electrocardiogram usually shows left

was subsequently confirmed at postmortem examination The patient died during an angiocardiographic examination

Tricuspid Atresia without Transposition of the Great Vessels This category of patients is also subdivided into two types (1) associated patent ductus and pulmonary atresia, (2) associated pulmonic stenosis and a 'slitlike' small ventricular septal defect

Figure 504 illustrates the case of a nine year old girl who had tricuspid atresia without transposition She was cyanotic, had slight clubbing of her fingers and toes, and had dyspnea on exertion On auscultation a grade III systolic murmur was heard along the left sternal border, loudest at the third left interspace The second sound was closely split. A third sound was present at the apex The diagnosis was established by angiocardiograms and cardiac catheterization.

TRANSPOSITION OF THE GREAT VESSELS (PULMONARY ARTERY AND AORTA)

Transposition of the great vessels is rarely encountered in adults because the great majority of patients with this anomaly die in infancy In fact few survive the first year of life This congenital defect accounts for a significant number of deaths among babies who die of congenital heart disease during the first months of life Cyanosis and clubbing variable in degree are present As a rule the cyanosis is quite evident In order for the patients to survive, shunts are necessary and these are usually in the form of septal defects (atrial and/or ventricular) or patent ductus A systolic murmur is usually present but some patients have no murmur

Transposition may be corrected or partially corrected by nature in rare cases For example a total anomalous venous drainage emptying

murs were present at the apex and base, and the second heart sound was slightly accentuated *Truncus arteriosus* was considered but there was close splitting of the second heart sound which aided in the clinical elimination of this possibility Based on the total cardiovascular evaluation, including angiocardiography, a diagnosis of transposition with partial anomalous venous drainage was made

TRUNCUS ARTERIOSUS

In this condition a common trunk arises from both ventricles A ventricular defect occupies the upper portion of the septum, or some

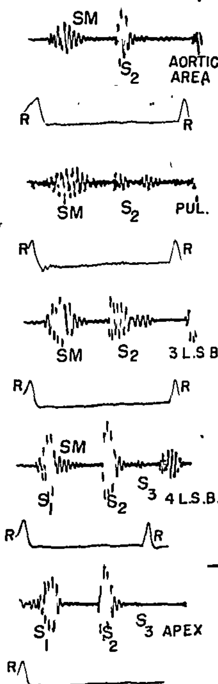
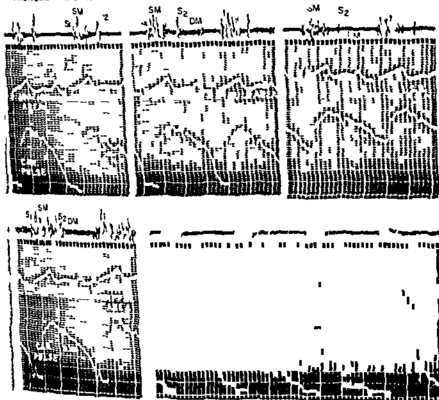


FIG. 504. 9 year old girl with tricuspid atresia and atrial and ventricular septal defects. Auscultation unremarkable except for faint grade III systolic murmur (SM) heard best along left sternal border. A third sound (S_3) heard along lower left sternal border and apex. Patient had dyspnea, cyanosis and slight clubbing of fingers and toes.

times the entire septum is absent. Cyanosis and/or clubbing of varying degrees are present, depending on the amount of blood supply to the lungs. With a larger pulmonary artery cyanosis is slight or undetected; with a small pulmonary artery it may be extreme. The pulmonary arteries may arise from each side of the single trunk, or variants may be seen where *only one* artery, or occasionally no pulmonary



TRUNCUS ARTERIOSUS — CONTINUOUS MURMUR BEST HEARD RT NIPPLE AREA and left sternal border

artery arises from the main trunk. In the latter case the lungs are entirely supplied by the bronchial arteries.

The murmur likewise is variable. It may be of a continuous machinery type or may have systolic and diastolic components which do not sound continuous. Sometimes the continuous murmur envelops the second sound in the same way as a typical patent ductus murmur. It may be heard best over the pulmonary area, and if this is the case may be confused with the murmur of patent ductus. A differentiating point is the finding of this murmur to the right of the sternum (at the second right interspace) *or over another area* of the chest

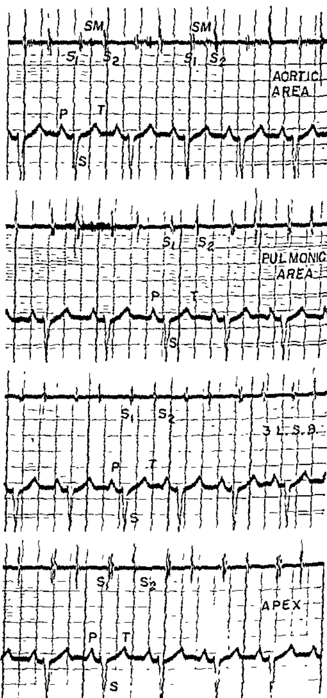
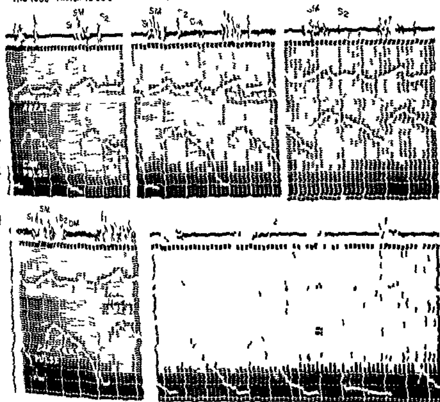


FIG. 505 A 17 year ... at vessels and ... Faint systolic ... cardiovascu-

times the entire septum is absent. Cyanosis and/or clubbing of vary- ... present depending on the amount of blood supply to ... ht or un ... The pul- monary arteries may arise from each side ... k, or vari- ants may be seen where only one artery, or occasionally no pulmonary



artery arises *from the main trunk* In the latter case the lungs are entirely suppl ...

... na
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the second sound *in the* same way as a typical patent ductus *murmur* It may be heard best over the pulmonary area and if this is the case may be confused with the murmur of patent ductus A differen- tiating point is the finding of this murmur to the right of the sternum (at the second right interspace) or over another area of the chest,

such as the third left or right interspaces. At times the murmur is not of the typical machinery type, and on careful auscultation the peak of the systolic murmur is heard in midsystole and therefore does not envelop the second sound (Fig. 506). It may sound "to-and-fro."

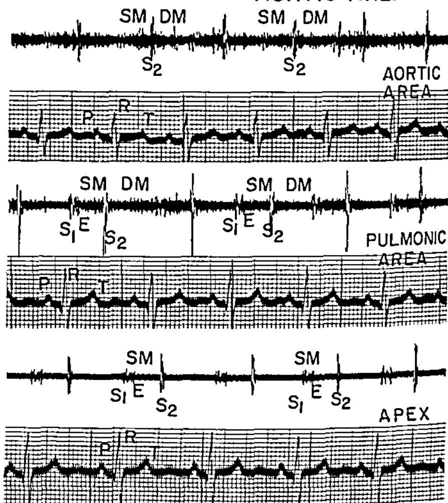
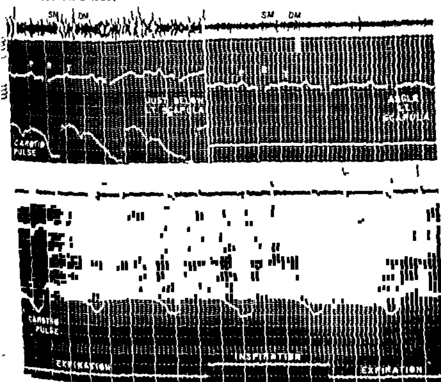


FIG. 507 Boy, age 7, with diagnosis of truncus arteriosus made by cardiac catheterization and angiocardiograms. Symptomatic; Slight cyanosis. Note continuous murmur with late systolic (SM) and early diastolic (DM) accentuation that enveloped second sound (S_2), loudest over aortic area (upper tracing); also well heard over pulmonic area (middle tracing). Note single second sound (S_2) in all areas. Also, early systolic ejection sound (E) shown in second and third tracings. Had been confused with patent ductus arteriosus.

As seen in Figure 506, and as pointed out earlier in the discussion on patent ductus, the finding of an atypical continuous type of murmur should always lead one to suspect something other than patent ductus. At least 95 per cent of patients with patent ductus are thereby ruled out. The truncus murmur is only systolic in some cases and is heard most frequently at the second left or second right interspaces, although occasionally it is located at the lower sternal borders or at

the apex. Rarely, no murmur has been present, although we personally
] ... a case The second sound is single (Figs
... The find
... ut a diag
nosis of truncus The second sound is ... , although
single, may at times be of longer duration than a fine, sharp single
sound Because of this it is sometimes confused with splitting
Frequently an early systolic sound (ejection sound) is heard at the



) (lower tracing) Second sound (S_2) was single No splitting with respiration.

ase and also at the apex (Figs 506, 507, 508) The first heart sound
variable and has no specific diagnostic significance Truncus arteri
sus should be suspected, therefore if there is a combination of a
murmur, continuous or systolic particularly if heard elsewhere than
ver the pulmonic area a single second sound an ejection sound
ariable cyanosis or clubbing electrocardiographic evidence of left,
ight or combined ventricular hypertrophy, x ray evidence of an
absence of the pulmonary artery segment together with the presence
of a common trunk, frequently to the right of the sternum Vascular
markings are dependent on the blood flow, angiocardiography is help

ful in identifying the condition, showing a main trunk from which pulmonary branches arise.

In general, the louder the murmur the slighter the degree of cyanosis. Adults are more likely to show a continuous murmur because of a better flow of blood to the lungs. Such patients have a better prognosis. In fact, patients with truncus whose condition allows only a

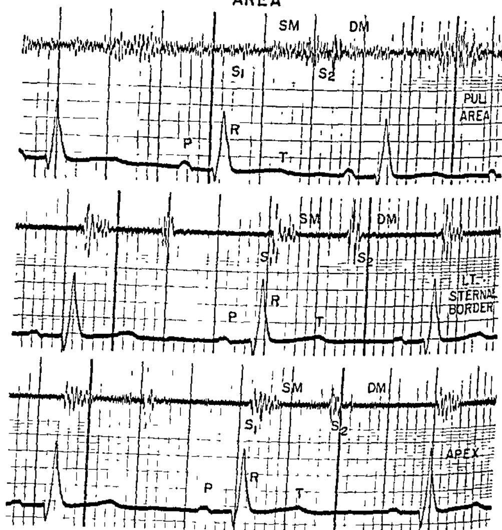
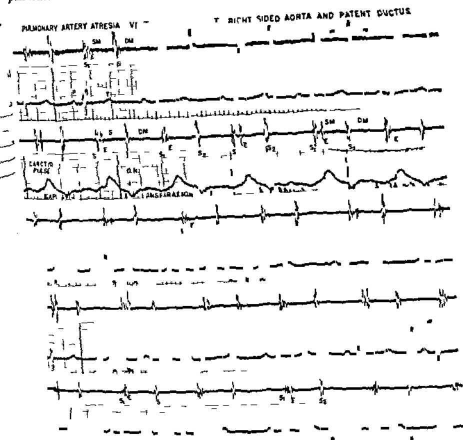


FIG. 509. Boy, age 8, with pulmonary atresia and ventricular septal defect (same patient as Fig. 482) Had cyanosis and a continuous murmur (SM–DM) heard best over pulmonary area (upper strip).

minimum blood flow to the lungs and who have a slight murmur generally die early in infancy.

Pseudotruncus. This condition is a variant of the typical tetralogy of Fallot in which the pulmonary valve or infundibular area is atretic. The bronchial arteries and/or a patent ductus supply arterial blood to the lungs. Cyanosis is regularly present, and the electrocardiographic findings are consistent with right ventricular enlargement. A

continuous murmur is common, as illustrated by the cases of the two patients shown in Figures 509 and 510 In one (Fig 509), the continuous murmur enveloped the second sound and was loudest over the pulmonic area In the other (Fig 510) the patient had a right-sided aortic arch, and the continuous murmur was loudest just beneath the right clavicle It was subsequently demonstrated that in addition to pulmonic atresia this patient had a patent ductus arising from the



ejection sound (E) in all areas

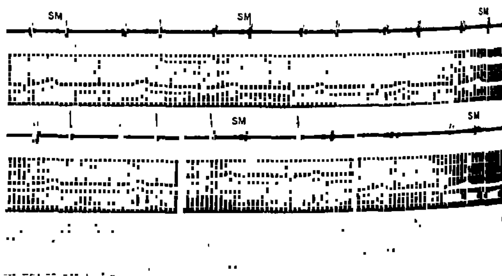
right-sided aorta This was the probable explanation for the continuous murmur heard best under the right clavicle A single second sound was present in both patients

CONGENITAL PULMONARY ARTERIOVENOUS FISTULA

This congenital connection between an artery and a vein occurring in the lung is a rare anomaly Pulmonary arterial blood which is

unsaturated is shunted into the venous side and arterial unsaturation results. With the larger shunts, cyanosis, clubbing of the fingers and toes and polycythemia may result. However, some patients do not show these signs, and the diagnosis is then more difficult. Approximately one-half of the patients with this congenital lesion have evidence of hemorrhagic telangiectasia (Rendu-Osler-Weber disease). The presence of the pulmonary lesion, therefore, may first be suspected by the finding of telangiectasia of the lips or oral mucosa, and such patients may have nosebleeds or symptoms or signs of gastrointestinal bleeding. An x-ray examination of the chest may show an abnormal shadow in the lung field at the periphery or in proximity to the heart. Angiocardiography is most useful in establishing the diagnosis by showing that the shadow is vascular.

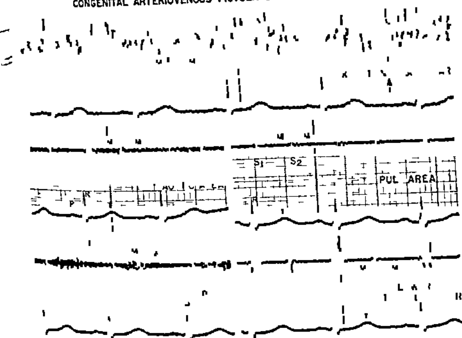
Auscultation may be of great value in diagnosis. A murmur may be



produced which can be continuous or only systolic. In our experience the majority of murmurs heard with this condition have been systolic (Fig. 511), although some are continuous. The finding of an unexplained murmur in an atypical area over the lung field, and, in particular, when there is no associated cardiac lesion, is an immediate clue. Sometimes with inspiration the murmur may be accentuated (Fig. 511), while in other patients the murmur may decrease. This decrease is probably due to the interposition of lung tissue coincident with inspiration. The increase with inspiration, on the other hand, could be related to increased blood flow. One or the other of these factors may predominate in individual cases. When a murmur is heard in the periphery of the lung or in the posterior lung field and is not associated with any cardiac auscultatory findings, the possibility of arteriovenous fistula must be seriously considered. On the other hand, some of these pulmonary lesions occur in close proximity to the heart

and may be misinterpreted
x rays or angiograms may l
obscured by cardiac or great vessel shadows
are helpful in eliminating this possible source of confusion Angio
cardiography is most valuable not only for confirming the diagnosis,
the po h lity of other similar pulmonary lesions

can occur from these fistulas and may cause death Other



matic.

of the patient s family should be examined for this anomaly because of its familial nature Confusion with congenital heart disease is most likely in those patients having cyanosis clubbing and a murmur in the precordial area Cardiac enlargement and cardiac decompensation may be present with some of the larger shunts

CONGENITAL ARTERIOVENOUS FISTULA OF THE CHEST WALL

A murmur had been present since birth in the four year old girl whose case is illustrated in Figure 512 Auscultation revealed the

murmur to be loud and continuous. Over the precordium it was louder at the aortic area than the pulmonic, but the sound was heard even better anteriorly over the right lower chest. Over the back, however, the source of the murmur became evident through the finding of a palpable thrill and a grade VI continuous murmur at the right upper

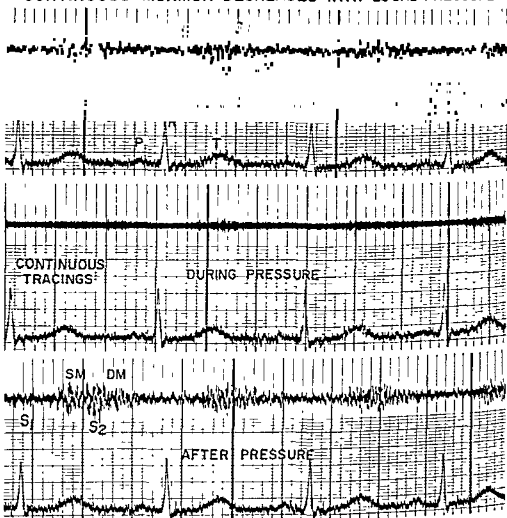


FIG. 513. Same patient as Fig. 512, showing effect of pressure over area in back. Upper tracing before pressure shows continuous murmur (SM–DM) which became fainter with local pressure (middle tracing) and louder again after release of pressure (bottom tracing).

interscapular region (Fig. 512). Pressure with the fingers in this area resulted in a striking decrease in the murmur's intensity, although it did not disappear (Fig. 513). The diagnosis of an arteriovenous fistula of the chest wall was made. No symptoms or signs of heart disease were present.

Another patient (Fig. 514) sought medical advice after his wife had detected a peculiar sound coming from his chest. A roaring noise was

evident and on auscultation, a continuous machinery type of murmur was heard Finger pressure over a well localized spot between the ribs caused the murmur to disappear, pointing to a diagnosis of an arteriovenous communication between the intercostal or internal mammary vessels Whether this represented a congenital or acquired lesion was not apparent However, there was no history of trauma.

In the case illustrated in Figure 515, a 22 year old woman had a ... murmur that was loudest over the pulmonic area. There ... in inten... with the ...thoscope caused a decrease in the murmur An operation for patent

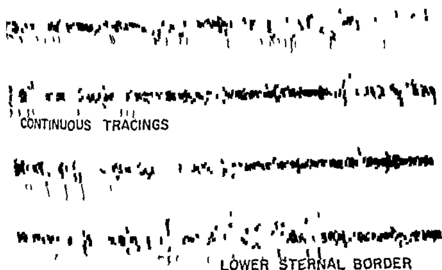


FIG. 514 A loud machinery murmur from arteriovenous fistula of chest wall Condition first noted by wife who heard strange noise emitting from patient's chest. (Phonocardiogram made from original tape recording)

ductus had been scheduled but was canceled because of these findings A cardiac catheterization was normal

It is clear from these cases that an arteriovenous fistula of the chest wall is often misdiagnosed or confused with cardiac lesions such as patent ductus pulmonary arteriovenous fistula or others No operations were performed in any of the cases just discussed, for the condition is regarded as essentially benign

CONGENITAL COMPLETE HEART BLOCK

A diagnosis of congenital complete heart block can be made with certainty if the block is found at birth or shortly thereafter In fact

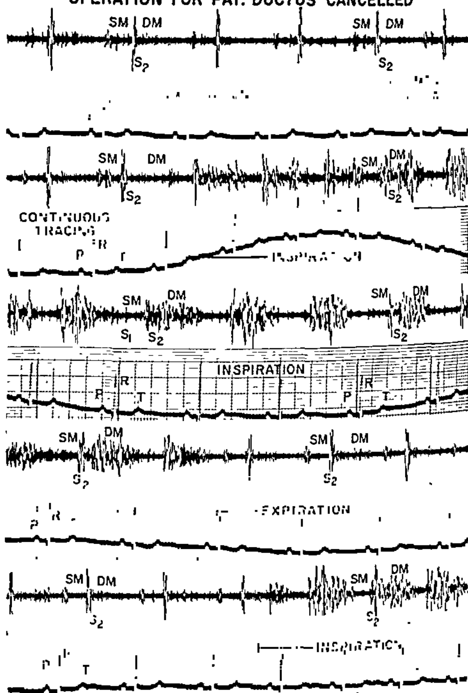
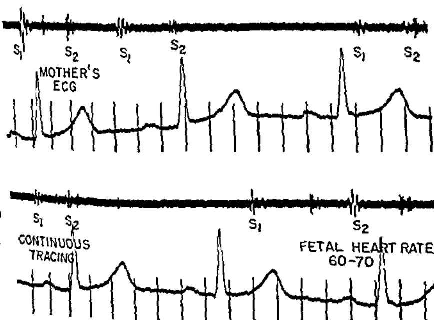


FIG. 515. A 22 year old woman with a conti[illegible] heard over pulmonic area. With expiration the [illegible] systole but without late systolic accentuati[illegible]

[illegible] phase. With pressure applied locally over area the murmur became much [illegible] Cardiac catheterization normal.

he diagnosis has been made in utero because of an abnormally slow fetal pulse rate Occasionally a changing intensity of the first fetal heart sound is noted in addition to the slow rate Figure 516 illustrates an example of this where the correct diagnosis of heart block was made one week before birth Many cases, however are overlooked for a number of years and then are discovered on routine physical examination. The diagnosis of congenital complete block can be made if it is discovered in a young patient with no history of coronary disease diphtheria, recent rheumatic fever or other infectious diseases



he auscultatory findings are essentially the same as in acquired heart block and include a slow rate, which generally averages 10 to 15 beats faster than in patients with acquired block The average ventricular rate in congenital complete heart block would range between 50 and 60 (Fig 517) This rate is more apt to fluctuate than in acquired block, and increases after exercise There is a characteristic changing intensity of the first heart sound Atrial sounds or even short atrial murmurs may be present and are particularly apt to occur shortly

CONTINUOUS MURMUR PUL. AREA INCREASES $\bar{c}$ INSPIRATION
PROBABLE A-V FISTULA CHEST WALL — SCHEDULED
OPERATION FOR PAT. DUCTUS CANCELLED

SM DM S_2 SM DM S_2

SM DM S_2 SM DM S_2

SM DM S_1 S_2 SM DM S_2

INSPIRATION P R T P R T

SM DM S_2 SM DM S_2

EXPIRATION

SM DM SM DM

phase. With pressure applied *locally* over area the *murmur* became much fainter. Cardiac catheterization normal.

short, ending after approximately two thirds of systole In the past the presence of a systolic murmur along the lower left sternal border and the apex was interpreted as indicating an associated ventricular septal defect. In the light of more recent knowledge and more accurate diagnostic procedures and follow up studies, this systolic murmur is now thought to be functional in the usual case (Figs 517 518, 519) Further support of this view is the rarity of complete heart block in

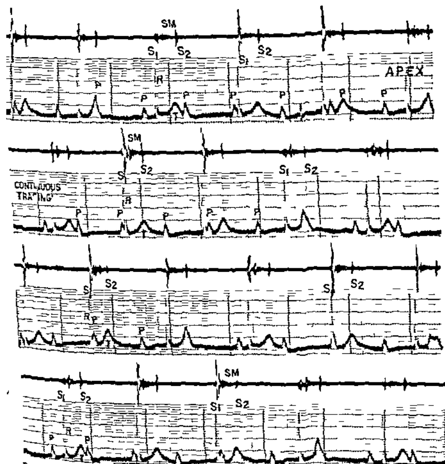


Fig. 519 Boy age 12 with slow heart rate (44) changing intensity of first sound (S_1) and short systolic murmur (SM)

cases of isolated ventricular septal defect Congenital complete heart block however does occur in association with other congenital defects of various types Figure 520 illustrates the case of a patient with pulmonic stenosis and complete heart block

The prognosis of uncomplicated congenital complete heart block is usually good In fact, in a number of instances it has first been detected on routine examination after patients have undergone very strenuous efforts such as vigorous sports or combat experience

CONGENITAL COMPLETE HEART (CONTINUOUS TRACING)

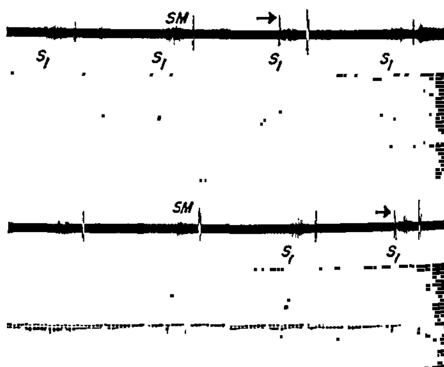
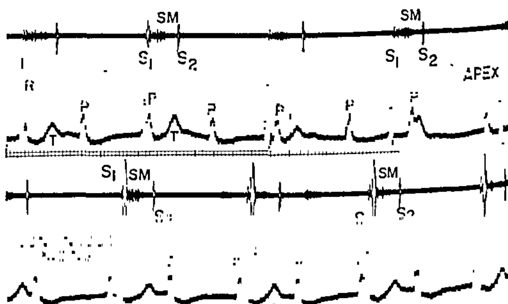


FIG. 517. Baby 9 days old. Showed slow regular rate of 50 with changing intensity of first sound (S_1) and grade III systolic murmur (SM) heard best over midprecordium. Loud first sounds indicated by arrows.

CONGENITAL COMPLETE HEART BLOCK



short, ending after approximately two thirds of systole In the past the presence of a systolic murmur along the lower left sternal border and the apex was interpreted as indicating an associated ventricular septal defect. In the light of more recent knowledge and more accurate diagnostic procedures and follow up studies, this systolic murmur is now thought to be functional in the usual case (Figs 517, 518, 519) Further support of this view is the rarity of complete heart block in

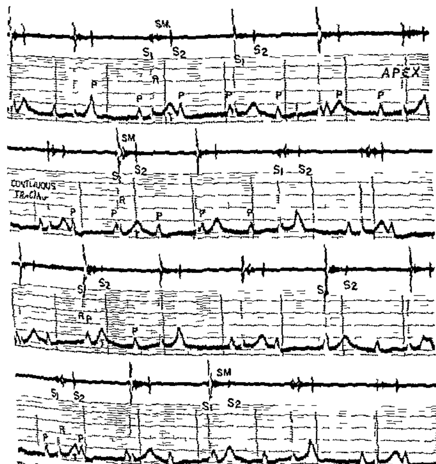


Fig 519 Boy age 12 with slow heart rate (44) changing intensity of first sound (S_1) and short systolic murmur (SM)

cases of isolated ventricular septal defect Congenital complete heart block however, does occur in association with other congenital defects of various types Figure 520 illustrates the case of a patient with pulmonic stenosis and complete heart block

The prognosis of uncomplicated congenital complete heart block is usually good In fact, in a number of instances it has first been detected on routine examination after patients have undergone very strenuous efforts, such as vigorous sports or combat experience

Syncopal episodes may occur but are unusual. Likewise, sudden death due to Adams-Stokes syncopal attacks is relatively rare. Although the prognosis generally is good, one cannot make predictions in the individual case, as evidenced by our recent observation of a young woman in her twenties who had congenital complete heart block. She developed Adams-Stokes syncope and died in one of these episodes. Previously she had the characteristic auscultatory findings of complete heart block, and in addition had a grade III systolic murmur that was

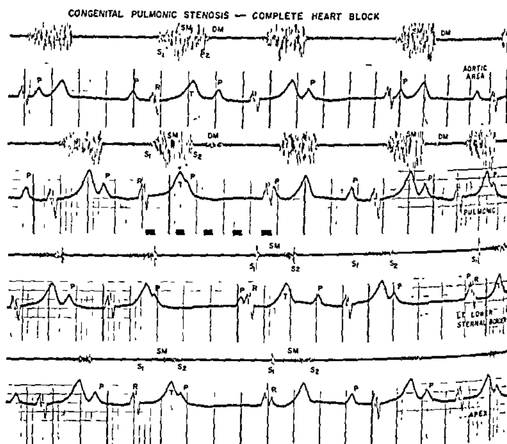


FIG. 520. Patient with congenital pulmonic stenosis and complete heart block. Systolic murmur (SM) loudest over pulmonic area (second strip). Also note diastolic murmur (DM) apparently related to atrial contraction (P on electrocardiogram); changing intensity of first sound (S_1) at left lower sternal border and apex (lower two tracings).

heard along the lower left sternal border and at the apex. At postmortem examination no associated congenital defect, such as ventricular septal defect, was present.

In some patients with congenital complete heart block the heart size is normal; in others it may appear to be on the outer limits of normal, or even slightly enlarged. On the other hand, with associated congenital defects, the heart may show significant enlargement. The outlook for these latter patients will generally depend upon the prognosis of the associated defect. It is of further interest that women

with congenital complete heart block as an isolated finding usually tolerate normal pregnancy without difficulty

CONGENITAL BUNDLE BRANCH BLOCK

As an isolated lesion bundle branch block right or left, can be present from birth but is unusual In itself it is unassociated with symptoms however when it is present it is usually associated with some other cardiac disease If bundle branch block is discovered in youth as an incidental finding in an otherwise normal individual it may well be regarded as congenital One cannot be certain of this, however since infections such as diphtheria pneumonia, etc, cannot

CONGENITAL RIGHT BUNDLE BRANCH BLOCK

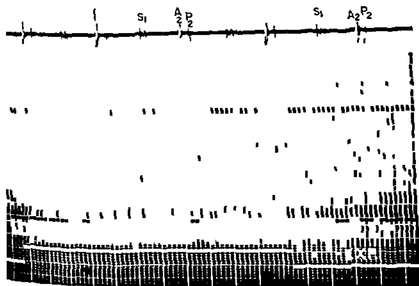


FIG. 521 Man age 21 with right bundle branch block thought to be congenital. Note splitting of second sound (A_2–P_2) which becomes wider on inspiration.

always be ruled out Figure 521 illustrates this problem This patient was diagnosed as having a congen ... ndle branch block There ... became still wider with ... s lesion, see Splitting of ... (page 47) and Bundle Branch Block (page 188))

IDIOPATHIC DILATATION OF THE PULMONARY ARTERY

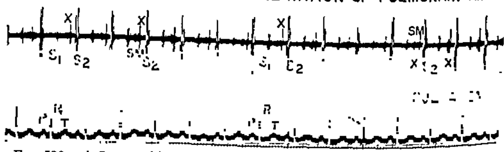
The pulmonary artery may show a definite enlargement that is frequently first discovered on a routine x ray examination of the chest. The physician must then rule out the possibility of congenital heart disease mitral stenosis or other causes of pulmonary artery enlargement.

IDIOPATHIC DILATATION OF PUL. ARTERY

X
SM
X
S_1 S_2
PUL AREA
P R T
S_1 X S_2
X
APEX
P R T

FIG 522. A 23 year old woman with idiopathic dilatation of the pulmonary artery. *Cardiac catheterization normal.* Had grade III systolic murmur (SM) over pulmonic area (upper strip). Systolic ejection sound heard over pulmonary area and also at apex. *Electrocardiogram normal* Patient asymptomatic.

SYSTOLIC SD. WITH IDIOPATHIC DILATATION OF PULMONARY ARTERY



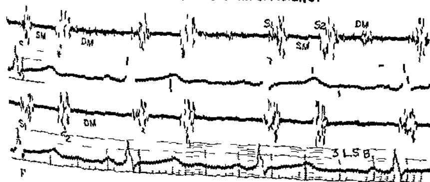
In a patient with an idiopathic dilatation of the pulmonary artery the electrocardiogram is normal and no other specific chamber enlargement of the heart is demonstrable Auscultation often discloses a characteristic sound in systole that is heard best over the pulmo- , area but which also may be heard at the apex This sound may be prominent as shown in Figures 522 and 523 In both patients the systolic sound was in about midsystole although in one (Fig 523) it was variable from middle to late systole A slight systolic murmur, grade I to III may be heard over the pulmonary area, and the second heart sound is normally split The degree of splitting increases in nor- with inspiration A total cardiovascular evaluation of the patients illustrated revealed no abnormality except the enlargement of the pulmonary artery segment and the auscultatory findings as mentioned Occasionally a diastolic murmur of pulmonary valve in ciency is heard

These patients should not be restricted in their activities but should be reassured and encouraged to lead a normal life Occasionally however patients with the so-called Marfan's syndrome may have dilatation of the pulmonary artery in which case the dilatation is presumably due to the medionecrosis More commonly Marfan s syndrome involves the aorta rather than the pulmonary artery

CONGENITAL PULMONARY VALVE INSUFFICIENCY

Insufficiency of the pulmonary valve as an isolated finding is an

valve cusps Figure 524 illustrates the case of a 36 year old lawyer



insufficiency Pulmonary artery segment moderately enlarged Electrocardiogram normal. Had faint grade II systolic murmur (SM) Patient asymptomatic

who has had a known murmur since the age of three. He is asymptomatic and was evaluated purely on the basis of the finding of a grade III early, blowing diastolic murmur that was heard best along the left sternal border. A grade I pulmonary systolic murmur was noted. No heart enlargement was evident fluoroscopically, but the pulmonary artery segment appeared to be moderately enlarged. There were no peripheral signs consistent with aortic insufficiency, and the electrocardiogram was normal. These findings were consistent with the diagnosis of a congenital pulmonary valve insufficiency that had produced no symptoms over a 36 year period.

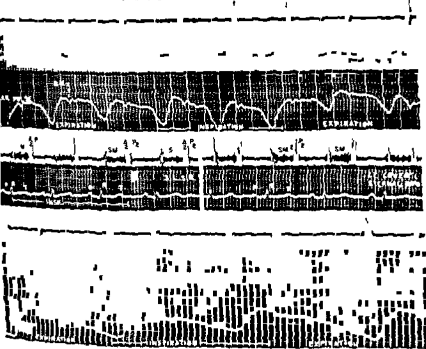
A postmortem examination was recently made on a patient who had had congenital malformation and hypoplasia of the pulmonary valve. He died at the age of 54, at which time he showed signs of right ventricular hypertrophy and pulmonary vascular arterial changes. Before death he had shown features consistent with cardiac decompensation. It is of interest, however, that patients may tolerate insufficiency of the pulmonary valve for many years without symptoms, and it is conceivable that a minimal leak of the pulmonary valve actually may never produce symptoms.

PRIMARY PULMONARY HYPERTENSION

Primary or idiopathic pulmonary hypertension occurs as a result of extensive proliferation of the intimae of the smaller pulmonary arteries, resulting in a decrease in the size of the arterial lumen, or even in its obliteration. Its cause is unknown and there are no associated congenital or acquired heart lesions. The resultant clinical picture is enlargement of the pulmonary artery; right ventricular enlargement, as evidenced by x-ray and electrocardiogram; cyanosis, which is peripheral in type; syncope; exertional dyspnea; precordial chest pain; and symptoms of congestive heart failure. Hemoptysis or arrhythmias may be present.

On auscultation a constant finding is an accentuated second sound over the pulmonary area (Fig. 525) similar to that already described in cases of pulmonary hypertension associated with the various congenital defects. The second pulmonic sound frequently can be palpated; a prominent atrial wave is seen in the jugular pulse; and a right ventricular lift or heave is felt along the left sternal edge, indicating right ventricular hypertrophy. The accentuated second heart sound is generally split, but closely so. Coincident with inspiration the degree of splitting frequently widens slightly in normal fashion. In our experience, a wide splitting of the second sound over the pulmonary area would be a point against a diagnosis of significant pulmonary hypertension. The absence of a significantly accentuated second sound in this area would also weigh against the diagnosis. The first heart sound at the apex is variable and appears to have no diagnostic significance. A prominent atrial sound or gallop may be

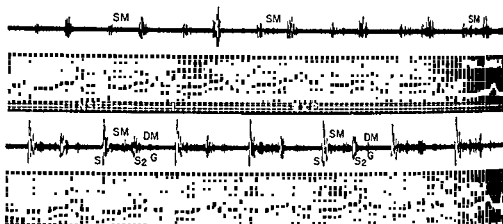
rd along the lower sternal border or apex, and in the presence of decompensation a ventricular diastolic gallop rhythm like- e may be evident. Over the pulmonary area a systolic murmur is mon, and this generally is of grade II to III intensity Loud sys c murmurs over this area are unusual in these patients At the ex a faint systolic murmur of grade I to III intensity may be pres (Fig 526) A frequent finding particularly in the more advanced ses, is a blowing diastolic murmur presumably due to insufficiency



reard in all areas, best along third left sternal border and left lower sternal border (middle strips) With inspiration a closely split second sound (A P) widened slightly with inspiration (lower tracing)

of the pulmonic valve In some the murmur is faint (grade I to grade II) while in others it may be grade IV As shown in Figure 527 the ... nal ... ole ... as in other cases of pulmonary hypertension (Figs 525 526) On x ray examination the lung fields at the periphery are clear

The outlook for patients with primary pulmonary hypertension is poor and no specific treatment has yet been effective Sudden death



PRIMARY PULMONARY HYPERTENSION

S1 S2 DM S2 DM S1 S2 V G DM APEX

FIG. 527. A 42 year old man with primary pulmonary hypertension. Note loud second sound (S_2) heard best over pulmonary area (upper strips) and early, blowing diastolic murmur (DM) of pulmonary valve insufficiency (second and third strips). At apex (lower strips) ventricular diastolic gallop (VG) and short diastolic murmur (DM) heard.

may occur but most of these patients have a progressive downhill
... ptoms of congestive heart failure

... r
... r

... unexplained, for autopsy revealed no ...
... lity, although the characteristic pulmonary arterial changes

... pertension. This was illustrated by a 42 year old patient ...
626 p 612) The choice in differential diagnosis was between ... y pulmonary hypertension or hypertension secondary to pul... monary emboli The patient was eventually incapacitated and had ... ncope, dyspnea hemoptysis chest pain and congestive heart failure ... stmortem examination revealed a right atrium filled with soft fri... ble clots and a main pulmonary artery partially occluded with an ... d organized thrombus An unusual auscultatory feature in this case ... as a widely split second sound This possibly could have been a ... elayed pulmonic valve closu... to the partial stenosis of the pul... monary artery from the thr... second sound was still loud ... ... ined by catheterization was 92 mm

EBSTEIN S ANOMALY OF THE TRICUSPID VALVE

Ebstein s anomaly is a congenital condition resulting in downward displacement of the tricuspid valve often with malformation As a result the upper ... nes a part of the right atrium The ... thin walled and grossly deformed ... natted against the right ventricular wall This results in insufficiency of the tricuspid

... blood from the right atrium shunts through a patent foramen ovale to the left atrium, cyanosis may result At times cyanosis may be present from birth, but at other times it is slow in developing In other cases no cyanosis whatsoever is evident One generally finds a systolic murmur varying from grade II to IV in intensity, that is heard best along the left sternal border and at the apex The murmur is sometimes loud harsh and associated with a palpable thrill These findings mimic pulmonic stenosis, with which Ebstein s anomaly may be confused At other times a diastolic rumble or a high pitched diastolic murmur is evident along the lower sternal border or at the apex Right bundle branch block is frequently noted on the electro

cardiogram, and prolongation of the P–R interval and large P waves consistent with right atrial hypertrophy may also be evident. Splitting of both heart sounds, and in particular the second heart sound, over the pulmonary area (possibly related to the right bundle branch block) is often heard. In addition, a ventricular or atrial diastolic gallop, or both, are common. These auscultatory findings are well illustrated in Figure 528 from a proven case of Ebstein's anomaly. Right bundle branch block was present; a grade III to IV systolic murmur and wide splitting of the second heart sound were noted. In addition, two sounds were present in diastole, which probably represented the ventricular diastolic gallop and the atrial gallop. The P–R interval was prolonged (0.23 second), resulting in a faint first heart sound In another instance of Ebstein's anomaly recently studied, the condition

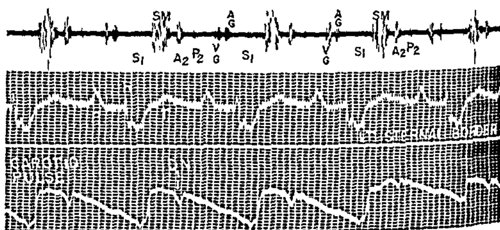


FIG. 5
(SM), wi
gallops

was confused with pericardial effusion because of the presence of a large, globular heart. This patient, a 53 year old mother, was essentially asymptomatic except for mild congestive heart failure. An angiocardiogram revealed the typical greatly enlarged right atrium of Ebstein's anomaly, and cardiac catheterization findings were consistent with this diagnosis. A grade III systolic murmur heard best along the lower sternal border was evident in this patient. No cyanosis was present.

At the present time no surgical treatment is available for patients with Ebstein's anomaly, and the possibility of confusing the condition with tetralogy of Fallot is thus particularly important. The x-ray findings in Ebstein's, however, are generally those of an enlarged and more globular type of heart and right atrial hypertrophy. The pulmonary artery segment is not enlarged, and vascular markings are usually normal or somewhat decreased.

DEXTROCARDIA

the more common form of dextrocardia the heart is on the right of the thorax and is a mirror image of the normal heart. It is ciated with situs inversus of the abdominal viscera Dextrocardia be the sole cardiac abnormality or may be associated with other

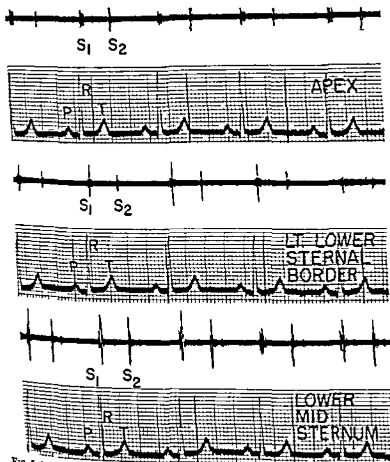


FIG. 5 9 A 26 year old woman with no heart disease. Her heart silhouette was more globular in shape than usual and occupied a middle portion of the chest. Note normal heart sounds (S_1 S_2) were heard best over lower midsternum (lower tracing)

defects. When there is dextrocardia without situs inversus other congenital cardiac lesions are the rule

Dextrocardia can be first suspected by palpating the maximum apical impulse over the right chest Auscultation in the uncomplicated case reveals heart ...

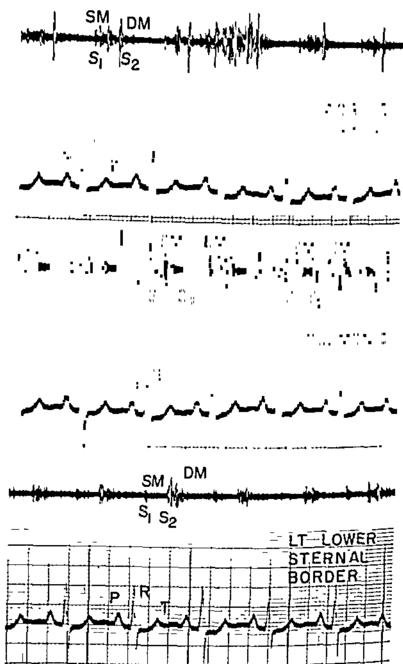


FIG. … murmur of pulmonar… systolic murmur (SM). … large aneurysm of pulmonary artery, and coarctation of the aorta. Cyanosis present.

and middle chest regions Rheumatic fever may involve the uncomplicated dextrocardia and result in the ordinary types of rheumatic valvular disease This can easily be overlooked unless one auscults on the right side. This possibility was illustrated by a patient observed at a teaching conference several years ago She had a history of rheumatic fever and classical symptoms of "tight" mitral stenosis A malar flush was noted on her cheeks The point of maximum apical impulse could not be located At first this was thought to be due to her obesity Over the customary areas of auscultation the heart sounds appeared somewhat faint but otherwise were not remarkable It happened, however, that in the patient previously presented to the conference, both sides of the chest were palpated simultaneously and the remark [illegible] the way we don t overlook dextrocardia " After be [illegible]

[illegible] mur- [illegible] augh

ter that filled the conference room

MESOCARDIA

The heart in this condition, which may be congenital, occupies the middle portion of the thorax The apex is often rotated rightward and anteriorly producing a more globular shape on the x ray film This may be the sole abnormality as illustrated in Figure 529 This was a 26 year old woman whose heart sounds were heard better over the lower midsternum than at the apex Mesocardia may be associated with other congenital cardiac defects and the auscultatory findings are dependent on the specific lesion In other instances, mesocardia is not congenital but represents a displacement of the heart rightward from various mechanical causes

MULTIPLE CONGENITAL HEART LESIONS

Congenital heart defects are often multiple For many of these patients the resulting disability is so great that death occurs in early infancy or childhood Some however survive into adolescence and adult life

Figure 530 illustrates an unusual example of multiple defects This patient an eight year old girl had a ventricular septal defect, coarcta

[illegible]

was heard best along the left sternal border

CHAPTER 4

Miscellaneous Auscultatory Findings

THERE are numerous other auscultatory findings of interest that are observed in a great variety of conditions. Most of these are well defined and well explained, while in others no satisfactory anatomic or pathologic background can be found. As will be seen, the findings in many of these conditions are quite specific and diagnostic, and for that reason intelligent auscultation is of vital importance.

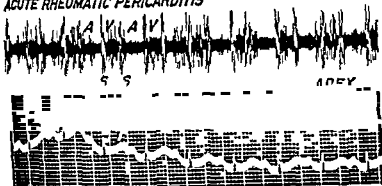
PERICARDIAL FRICTION

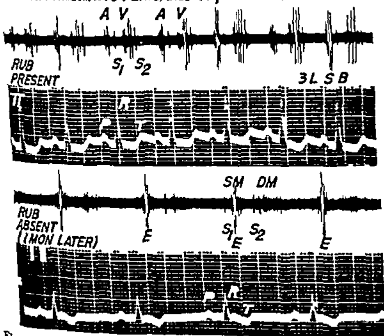
The detection of a pericardial friction is often the only means by which the diagnosis of acute pericarditis can be made. This is almost always a secondary complication of some other underlying disease. It is associated for the most part with the following conditions: rheumatic infection (Fig. 531), uremia (Fig. 532), tuberculosis, pneumonia (Fig. 533), acute coronary thrombosis (Fig. 534), general sepsis, virus infections (Fig. 533) and as a terminal phenomenon in cachectic conditions. There is also a form of acute pericarditis that may resemble acute coronary thrombosis or rheumatic pericarditis. This apparently runs a benign course, without subsequent complications, and may actually belong to the group of virus infections.

The pericardial friction which is practically pathognomonic of acute pericarditis is characteristically a scratching or grating to-and-fro sound. It appears to be dissociated from the first and second heart sounds, and when loud seems to be close to the ear. It may be sharply localized in a small area over the precordium, but often is loud enough to be heard well all over the left anterior chest. When faint it may become more clearly defined with the patient in the upright position. It is most frequently detected between the apex and the left sternal border, or even over the xiphoid area. Friction rubs often have high

[lo]w components and for this reason are generally heard best

[resp]iration frequently alters the intensity As a rule, there is an increase [w]ith normal inspiration (Fig 535) Although in its classical form





Fi[g] ... systolic (SM) and diastolic (DM) murmurs were heard Note ejection sound (E) of aortic insufficiency

there are systolic and diastolic components, there are rare instance in which only a systolic scratching sound will be audible. This ma become more typical some hours or days later. The pericardial fric tion may be quite transient and disappear in several hours, thoug generally it lasts for days.

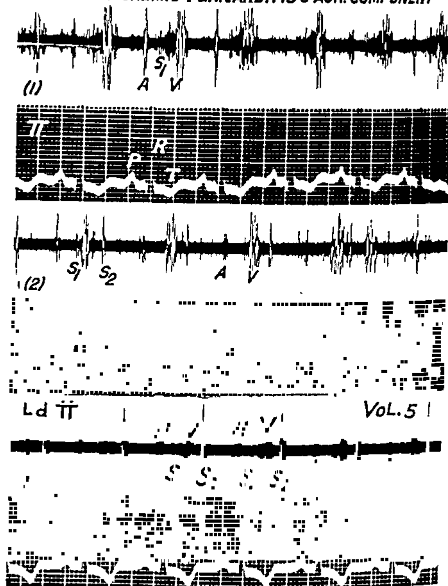


FIG. 532. Note systolic (V) and diastolic components (A) of pericardial friction rub in all three cases of terminal uremic pericarditis. Cases 1 and 3 were examined post mortem and showed extensive fibrinous pericarditis.

The friction sound does not always have a coarse grating quality. When this is the case, and because the sound is systolic and diastolic in time, it may closely resemble the to-and-fro murmur of aortic valvular disease. This differentiation is particularly difficult at times because in rheumatic heart disease acute pericarditis is more often

with aortic valvular disease than with mitral stenosis it is blowing rather than grating in quality, it resembles the in aortic insufficiency, and final judgment will have to be delayed If it disappears entirely, one can be *fairly certain that it was* a pericardial friction If it persists and remains unchanged, one can conclude that it was due to aortic valvular disease, for a friction does

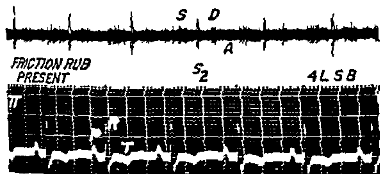
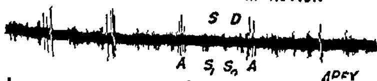




FIG. 5?3 Woman age 44 had pericardial friction rub associated with atypical pneumonia. Note systolic (S) and diastolic (D) components and a separate atrial component (A). Lower tracing shows normal sounds 14 months later



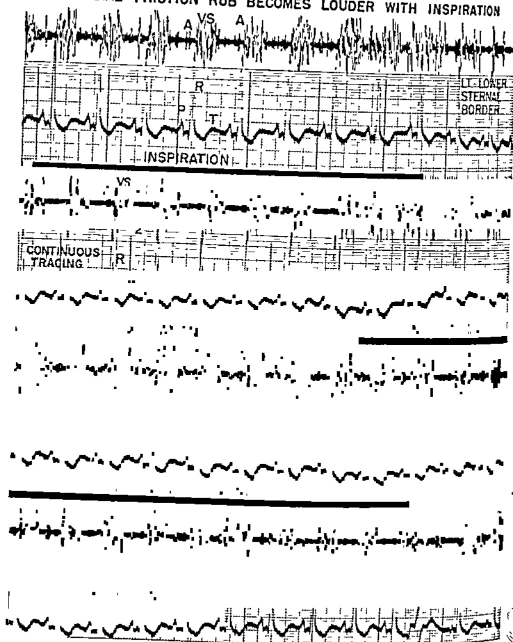
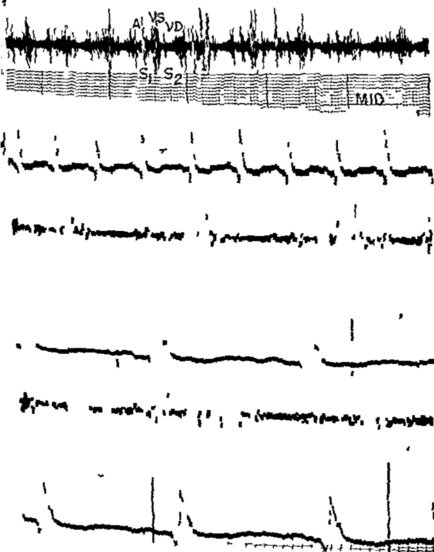


FIG. 535. Man, age 46, with uremic pericarditis. Had pericardial friction rub ... by atrial systole (A), ventricular systole (VS) and ... triking increase in friction rub coincident with ... component of rub (VD) led some observers to ... mortem examination revealed no valvular deformity.

not remain indefinitely. There are instances in which the grating component disappears, with the classical to-and-fro murmur of aortic insufficiency persisting. Under these circumstances, one would be justified in making the diagnosis of both conditions. In the early stages the friction may be so loud as to mask additional murmurs of chronic valvular disease. A somewhat similar problem arose in the case illustrated in Figure 535, a 46 year old man with uremia whose diastolic

blood pressure had been observed to drop A loud friction rub was present with a high pitched, blowing diastolic component. This caused some to make the diagnosis of acute aortic insufficiency on the basis of aortic valve rupture due to bacterial endocarditis A ventricular diastolic gallop and pulsus alternans were also present The patient died several weeks later and postmortem examination revealed acute fibrinous pericarditis but no associated valve deformity This case also illustrates the poor prognosis associated with the occurrence of a friction rub with uremia, death generally occurring within a few weeks



strip). A continuous tracing taken at the same area at faster speed (lower two strips) also illustrates three components of friction rub

As a rule pericardial friction rubs have two or three components. Various combinations can be present. The majority appear to have two components: atrial systole producing a presystolic rub, followed closely by ventricular systole producing a systolic rub. This close sequence accounts for the sound's simulating the rubbing of surfaces together (such as sandpaper on wood, creaking of leather, etc.) rather than systolic and diastolic murmurs. However, some frictions have two components made by ventricular systole and diastole. Here the diastolic component may be early and/or mid-diastolic. An atrial component may be present and then may result in a rub with three parts (Fig. 536). A systolic component alone is rare, except very transiently. It is generally good practice not to regard as a friction a sound with only a systolic component. More likely it will prove to be a scratchy systolic murmur that will persist. This is well illustrated by the case of a 74 year old woman. Because of a scratchy pulmonic systolic murmur of this type, a friction rub has been erroneously diagnosed on three separate occasions. This murmur has remained unchanged for at least eight years, which is as long as she has been under our observation.

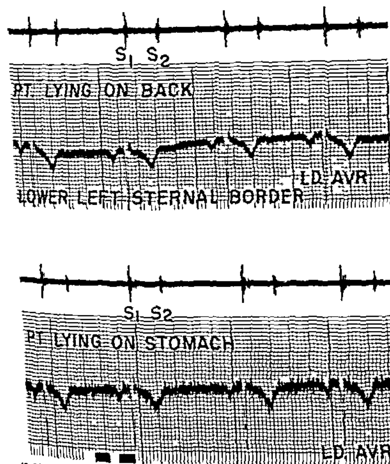
The movement of the ventricles during systole and diastole can account for the to-and-fro element of the friction rub. It is now quite well established that atrial systole may produce an additional friction sound when pericarditis is present (Figs. 531 to 536). In fact, there are instances in which the third portion of the friction sound due to atrial systole has been observed to disappear as atrial fibrillation set in and the atria ceased contracting.

PERICARDIAL EFFUSION

Both heart sounds may be markedly diminished in the presence of pericardial fluid. This is particularly true of a large effusion. On the other hand, with a moderate effusion the heart sounds may be of normal intensity. The x-ray examination may be helpful in diagnosing a large effusion, but not a small one. The electrocardiogram merely shows complexes of low amplitude with large effusions, but otherwise is of no diagnostic help. In doubtful cases, angiocardiography is sometimes resorted to; pericardial aspiration may be attempted, or a direct operative exploration is performed.

A useful sign in some cases has been the effect of the patient's position on the intensity of heart sounds. In normal individuals, as shown in Figure 537, heart sounds remain the same or become louder when the patient turns on his stomach. Auscultation is then easier and more convenient if the patient props himself up on his elbows. In these positions, the heart is in closer proximity to the chest wall. On the other hand, in some patients with pericardial effusion, the heart sounds become fainter or remain the same after these changes in

. . . ho d agnosis of peri- by the usual typical signs ericardial effusion, and this diagnos s — at a postmor- examination when a tumor of the pericardium was found



IG. 537 Young physician with no heart disease First (S_1) and second (S_2) heart sounds remained unchanged on change of position

The sign was most useful in the case of a 45 year old woman with heumatic mitral stenosis and insufficiency aortic stenosis and insuf iciency and tricuspid insufficiency (Fig 539) The heart was en

inconclusive On turning the patient from her back to her abdomen

the heart sounds and murmurs were noted on the phonocardiogram definitely to decrease. Her condition continued to deteriorate, and one week later the heart by x-ray had significantly increased in size and the silhouette was now suggestive of pericardial effusion. Under direct vision in the operating room approximately 1,000 cc. of bloody fluid was aspirated from the pericardial cavity. Following this her condition improved, and two weeks later, as shown in the phonocardiogram, the effect of position on her heart sounds and murmurs was again tested. This time, there was a definite increase in both sounds and murmurs coincident with turning.

This auscultatory evidence of pericardial effusion is variable, how-

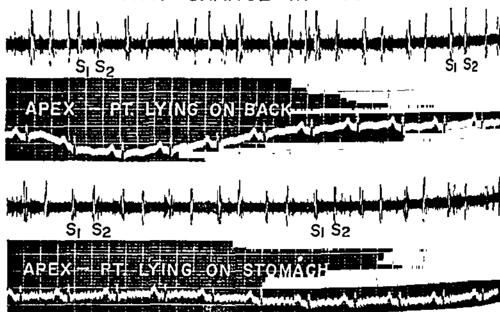


FIG. 538. Patient w pected (subsequently co position from lying on

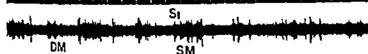
ever. We have observed that patients having large effusions may show little change with position. It is postulated that, with the more moderate amounts of fluid, coincident with change in the patient's position from the back to the abdomen, more fluid is interposed between the heart and the stethoscope, thereby decreasing sounds and murmurs. On the other hand, with the larger effusions, the pericardial sac being already greatly distended, there may be little change; in some instances even an increase has been noted. In general, the diagnosis of large effusions is not difficult.

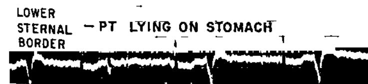
An example of little effect from change in position is shown in Figure 540. Here the heart sounds are faint in both positions, and the electrocardiogram shows slightly low voltage. Likewise, with smaller effusions there may be little change in intensi of the murmur with

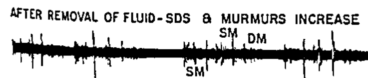
PERICARDIAL FLUID – DECREASE IN SOUNDS AND MURMUR WITH CHANGE IN POSITION



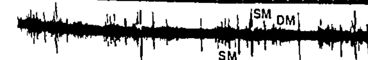












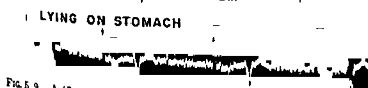


FIG. 5 9 A 45 year ... itral valvular lesions ... se in sounds systolic ... om back to stomach ... of bloody fluid from ... o effusion present. Sounds and murmurs are increased with ... anged position.

the heart sounds and murmurs were noted on the phonocardiogram definitely to decrease. Her condition continued to deteriorate, and one week later the heart by x-ray had significantly increased in size and the silhouette was now suggestive of pericardial effusion. Under direct vision in the operating room approximately 1,000 cc. of bloody fluid was aspirated from the pericardial cavity. Following this her condition improved, and two weeks later, as shown in the phonocardiogram, the effect of position on her heart sounds and murmurs was again tested. This time, there was a definite increase in both sounds and murmurs coincident with turning.

This auscultatory evidence of pericardial effusion is variable, however. We have observed that patients having large effusions may show little change with position. It is postulated that, with the more moderate amounts of fluid, coincident with change in the patient's position from the back to the abdomen, more fluid is interposed between the heart and the stethoscope, thereby decreasing sounds and murmurs. On the other hand, with the larger effusions, the pericardial sac being already greatly distended, there may be little change; in some instances even an increase has been noted. In general, the diagnosis of large effusions is not difficult.

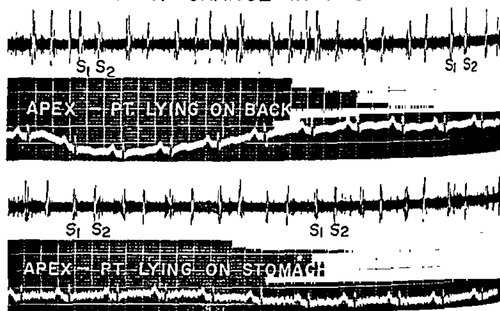


FIG. 538. Patient w ... pected (subsequently co ... position from lying on ...

An example of little effect from change in position is shown in Figure 540. Here the heart sounds are faint in both positions, and the electrocardiogram shows slightly low voltage. Likewise, with smaller effusions there may be little change in intensi of the murmur with

stenosis with a diastolic murmur This murmur, however, did not have the low frequency quality that is so characteristic of mitral stenosis On very rare occasions mitral stenosis may coexist with

PERICARDIAL KNOCK — CONSTRICTIVE PERICARDITIS

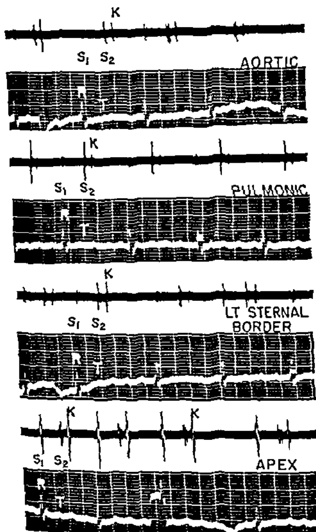


FIG 541 Woman age 53 with constrictive pericarditis Incapacitated with generalized anasarca. Note prominent pericardial knock sound (K) heard over precordium, best at apex (lower tracing) Pericardial calcification demonstrated on oblique and lateral x rays.

pericardial constriction The detection of this early diastolic sound or knock in a patient who does not have the customary signs of coronary hypertensive or rheumatic valvular disease should always make one suspect the existence of pericardial constriction

Tuberculosis is responsible for many cases of constrictive peri

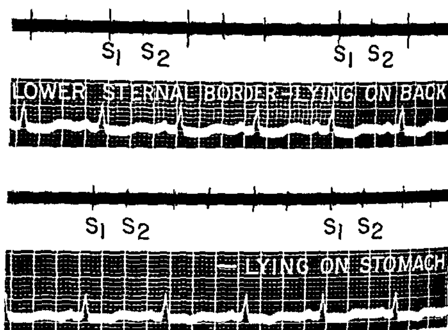


FIG. 540. Woman with pericardial effusion showing no significant change in hear sounds (S_1, S_2) on change in position. Both first (S_1) and second (S_2) sounds faint i both positions.

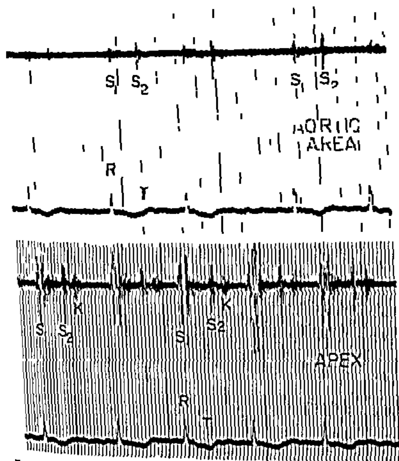
change of position. A false positive sign may be elicited in the presence of pleural fluid, which in some cases may cause a decrease in the intensity of heart sounds when the patient lies on his stomach. At present, this sign appears to be of value in selected cases.

CONSTRICTIVE PERICARDITIS

A sound that occurs early in diastole is a common finding in patients with constrictive pericarditis. It may be present with or without calcification of the pericardium. The term "pericardial knock" seems appropriate, for this sound apparently is produced in the rapid filling phase of ventricular diastole. The extra sound is a filling sound, as the constricting pericardium acts as a restricting shell, preventing the usual relaxation of the ventricles in diastole. It occurs in the early part of diastole at about the same time as, or slightly after, the opening snap of mitral stenosis. In fact, constrictive pericarditis often simulates, or is confused with, mitral stenosis. The typical rumble of mitral stenosis, however, is not present, although we have seen a case of constrictive pericarditis where the constriction involved the atrioventricular portion of the left ventricle, producing a relative type of

rovement from the surgery The pericardial knock sound in early iastole may show considerable variation in intensity and number of omponents (Figs 544 545 546) and also may vary with respiration Fig 547) The patient whose case is illustrated in Figures 544 and 545 had had his first operation fifteen years previously He obtained

PERICARDIAL KNOCK SOUND
CONSTRICTIVE PERICARDITIS



Note pericardial
also present in ad
surgery provement followed

some improvement, but subsequently developed gradual progressive cardiac decompensation On a second attempt at operation (at the age of 54) the constriction was still present and the patient died shortly afterward A systolic murmur had also been present but no diastolic rumble was heard In fact, during his several hospitalizations a number of observers first diagnosed the condition as mitral stenosis Atrial fibrillation and cardiomegaly were also present

carditis. Figure 541 shows a typical example of pericardial knock with constrictive pericarditis. The sound was heard best at the apex but also over the entire precordium—even over the aortic and pulmonic areas. This patient, a 50 year old woman, was completely incapacitated. She showed typical venous engorgement, hydrothorax, hepatomegaly, ascites and generalized anasarca. A prominent pericardial knock sound was present, but there was no diastolic rumble, and these findings provided the first clue to the correct diagnosis. Five years

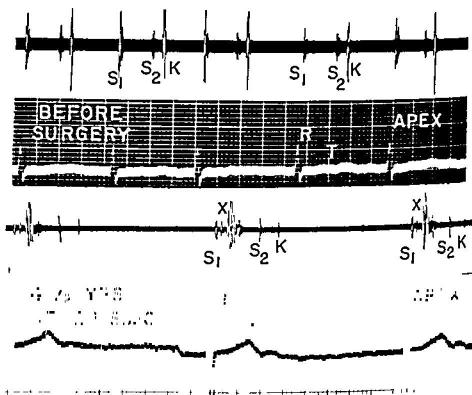


FIG. 542 Same patient as Fig. 541 before and after surgery. Obtained excellent symptomatic relief following operation. Before surgery note loud pericardial knock sound (K) in addition to first (S_1) and second (S_2) sounds. Four and one-half years after surgery (lower tracing) knock sound (K) was faint, occurring later after second sound (S_2) than before operation. In addition, systolic sounds (X) also present.

after her operation the patient was asymptomatic and able to carry on her work with no difficulty (Fig. 542).

The pericardial knock sound may be single and sharp, as it was in the case just described, or it may be of longer duration, as illustrated in Figure 543. The latter patient, a 74 year old woman, had this typical pericardial sound. An apical diastolic thrust, which is frequently palpated in these patients, was also noted. Calcified pericardial constriction was present. This patient was the oldest in our records who had this kind of operation, and she obtained good im-

surgery, occurring at a later interval than before surgery It is often at the time of a ventricular diastolic gallop or a normal third heart sound Whether this sound is related to the ventricular gallop of heart failure or to the remaining pericardial adhesions is not altogether clear It is well known that many patients who undergo surgery for constrictive pericarditis often wait for months, or even a year, before obtaining maximum benefit. An element of myocardial failure cer

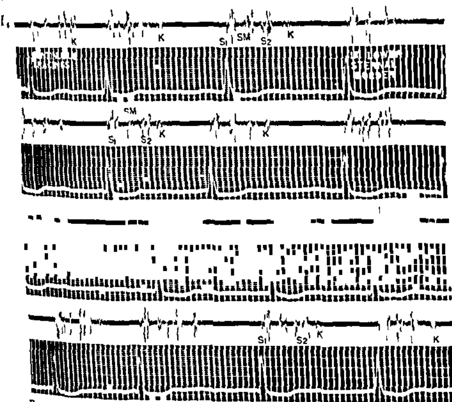


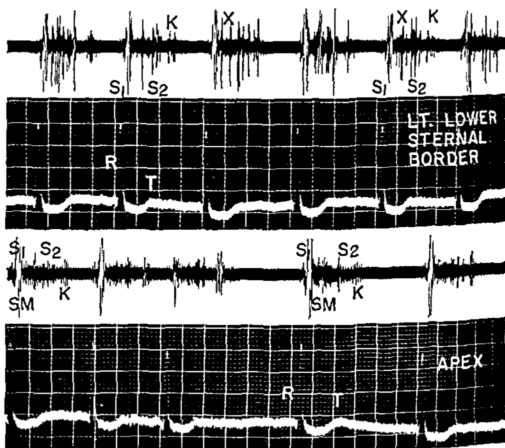
FIG. 545 Same patient as Fig 544 Taken at fast speed showing variation of knock sounds (K)

tainly exists in some of these patients Figure 542 represents a faint but definite pericardial knock in a patient with constrictive pericarditis following surgery

DIASTOLIC SOUND WITH CALCIFIC ADHESIVE PERICARDITIS (NOT CONSTRICTIVE) AND MITRAL INSUFFICIENCY (PERICARDIAL KNOCK SOUND)

Figure 549 illustrates the case of a 32 year old woman who had rheumatic heart disease with predominant mitral insufficiency X ray films revealed typical calcific pericarditis There was no evidence of

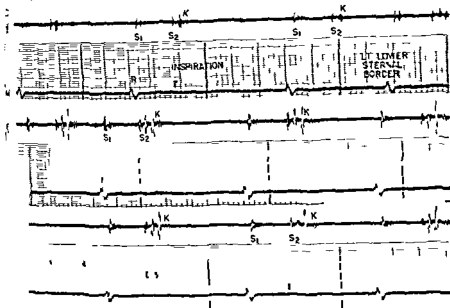
Continuous tracings taken at a fast speed along the lower sternal border show variations of the pericardial knock sound, diminishing on inspiration and becoming louder on expiration (Fig. 547). In this patient, the sounds were heard best at the apex and along the lower sternal border, but also could be heard over the base of the heart.



striction.

Atrial fibrillation was also present. Figure 548 illustrates the case of a 53 year old woman with advanced congestive failure who was incapacitated because of constrictive pericarditis. Her pericardial knock sound was composed of more than a single component and became louder with inspiration. Prompt improvement followed the operation.

The sounds may persist after surgery, although to a lesser degree, or may be absent. Sometimes a diastolic sound is present following



lowing several
Also note in

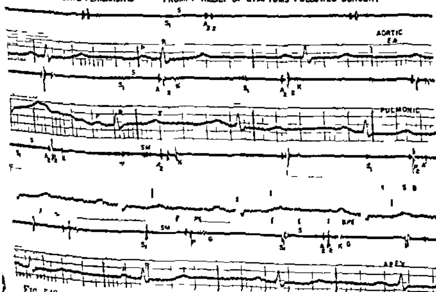


FIG. 548 A 53 year old woman with typical clinical features of constrictive

constriction, but a sound was heard in early diastole that was similar in mechanism, we think, to that of the knock of constrictive pericarditis. In this case it was postulated that the calcific pericardium shell, even though no constriction was present, acted as a rigid and

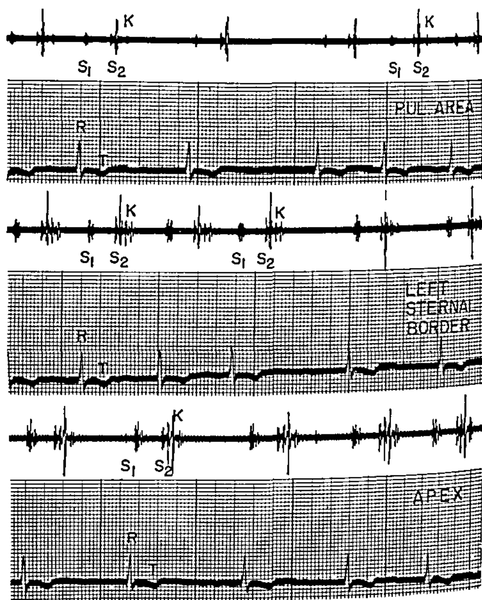
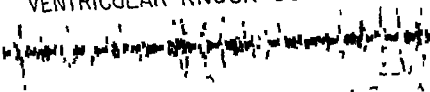
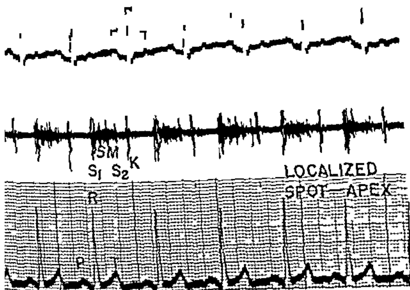


FIG. 546. Patient with constrictive pericarditis showing pericardial knock sounds (K) heard best at apex and left sternal border (lower two tracings).

abrupt block during the rapid diastolic filling phase of the ventricle. The diastolic sound was associated with a palpable impulse; it also waxed and waned with inspiration. In addition, this patient had a grade IV systolic murmur of mitral insufficiency.

2 PATIENTS WITH CARDIOMEGALY & VENTRICULAR KNOCK SOUNDS

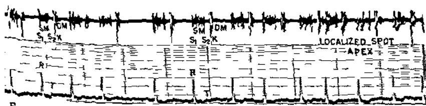




: 500 Upper tracing Man age 30 with severe aortic insufficiency and ex e left ventricular enlargement. Loud knock sound (K) in diastole occurred ltaneously with palpable impact at apex Systolic (SM) and diastolic (DM) murs

x a loud knock sound (K) was heard at the same time an apical impulse was pated. Severe cardiomegaly present

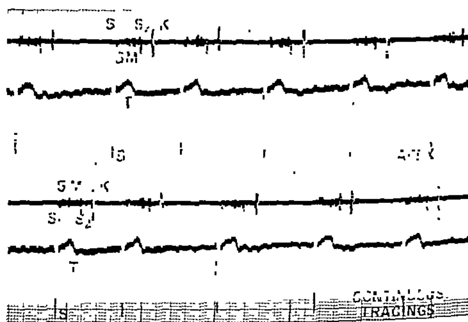
VENTRICULAR KNOCK SOUND — CARDIOMEGALY —



~ bu nas. Diastolic murmur (DM) also pre ent Knock sound (K) was loudest at localized spot at apex where prominent palpable lift occurred simultaneously

VENTRICULAR KNOCK SOUND IN DIASTOLE

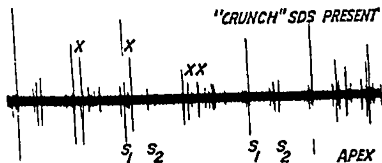
In patients with marked cardiac enlargement, particularly those with rheumatic heart disease and predominant mitral insufficiency and some element of stenosis, or combined lesions, and also in patients having severe aortic insufficiency with extreme degrees of left ventricular enlargement, a loud sound in early diastole may be heard. It is usually present over a localized spot at the apex at the point of maximum impulse and is also felt as a diastolic thrust in early diastole. We have termed this the "ventricular knock sound," feeling that it is produced by the impact of an enlarged ventricle against the

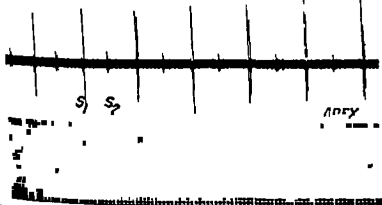


early pericardial knock sound (K) which varied in intensity. At times it was louder than either first (S_1) or second (S_2) heart sounds.

anterior chest wall in early diastole. This sound may be louder than even the first or second heart sound and is often misinterpreted as an opening snap or gallop. The mechanism of its production is thought to be analogous to that of the pericardial knock sound of constrictive pericarditis. Whereas the constrictive pericardial shell is responsible in constrictive pericarditis, the chest wall is the restricting structure with marked cardiac enlargement. The sound occurs later than the opening snap of mitral stenosis and is generally slightly later than the pericardial knock. It has the approximate timing of a ventricular diastolic gallop or normal third heart sound. We prefer the term "ventricular knock" because this sound is different from that of a gallop,

tesis. It has occurred after severe coughing episodes It has also occurred in underwater swimmers during World War II (frog men) When it occurs suddenly, it may produce symptoms strongly resembling acute coronary thrombosis (Fig 552) The main diagnos-





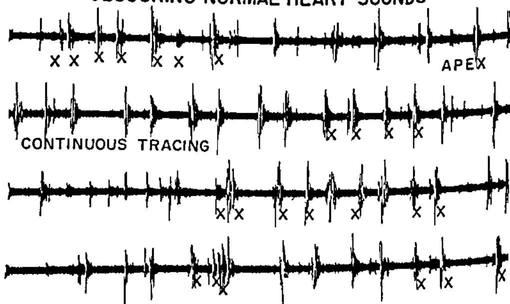
tic sign is elicited by auscultation One may hear bizarre crunching sounds over the precordium, mainly during systole and to a lesser extent in diastole (Figs 553 554) The sounds have been aptly de scribed as analogous to walking on crunchy snow They are more likely to be heard near the apex of the heart and at times become

which is usually much fainter; and, although a gallop can frequently be palpated, the ventricular knock produces a more striking impact.

Figures 550 and 551 illustrate this knock sound. The first patient in Figure 550 had marked cardiomegaly, particularly of the left ventricle, associated with severe aortic insufficiency. The knock sound was palpated and heard at the anterior axillary line. The second patient (Fig. 550) had cardiomegaly with rheumatic heart disease, severe mitral insufficiency and a giant left atrium. Slight mitral stenosis and aortic stenosis were also present. Another patient (Fig. 551) had extreme cardiomegaly from rheumatic heart disease, predominant mitral insufficiency and a giant left atrium. Note the pansystolic murmur of mitral insufficiency and the prominent, loud ventricular knock heard in this latter patient.

ACUTE MEDIASTINAL EMPHYSEMA (HAMMAN'S DISEASE)

There is an uncommon condition in which air suddenly infiltrates the anterior mediastinal tissue from the neighboring lung. This may occur spontaneously or after certain surgical operations involving the pleura or mediastinum. It is fairly common, however, following cardiac surgery, usually lasting hours, but occasionally lasting a day or two. Perforation of the esophagus should always be kept in mind as a cause. Sometimes it may follow pneumoperitoneum for treatment of tuberculosis, and occasionally it is a complication after a thoracen-



comes in paroxysmal form and may produce few, if any, symptoms

ons of the diaphragm It also gives rise to peculiar sounds that can be heard all over the chest The possibility that the condition may be present will most often occur to the physician on hearing peculiar sounds during auscultation of the heart. The sounds are usually low frequency and thus may be better appreciated using light pressure with the bell of the stethoscope The first time such a patient was observed by us, it seemed that one was listening to two different hearts within the chest. In the case illustrated in Figure 556, there was evi-

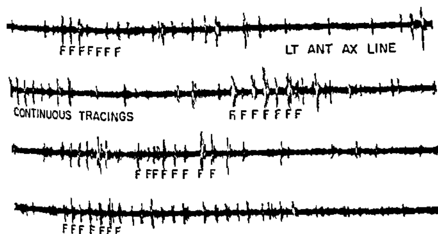
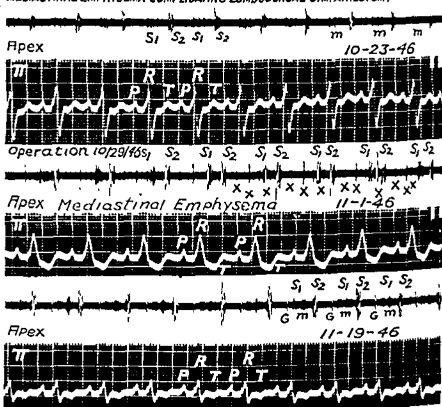


FIG. 556 Patient in a terminal state Had diaphragmatic flutter Flutter sounds (F) were clearly audible and louder than the normal heart sounds.

dence of rheumatic valvular disease However additional sounds were prominent that before the true nature of the problem was deter ined the observers thought that atrial fibrillation was present On more careful study it quickly became evident that fairly regular ounds entirely independent of the pulse were clearly audible in the ight axilla far removed from the heart Moreover, on auscultation ver the carotid arteries the real heart sounds could be heard and ounted at a normal rate of 75 It was then obvious that there were wo origins of the various sounds heard over the chest one from the eart itself and the other from some other source possibly the dia phragm The latter mechanism was confirmed by fluoroscopic exam ination Although these diaphragmatic contractions came and went without obvious cause they were present at times even during sleep The cause of this condition is still obscure

audible only when the patient lies in the left lateral position. Sometimes they are quite loud and may be heard without a stethoscope even at a distance of several feet. The patient himself is occasionally aware of these peculiar sounds and to bring them out, can assume a certain position or phase of respiration (commonly breath held in inspiration or with a Valsalva maneuver). The detection of air in the pleural cavity and sometimes in the anterior mediastinum on x-ray



anterior mediastinum. Lower tracing, 18 days later, no "crunch" sound. Note atrial gallop (G) and systolic murmur (M)

examination may aid in the diagnosis. The electrocardiogram may be normal, but occasionally shows T wave and minor S–T segment changes. Recovery is likely to be complete without any special medication, although recurrences occasionally take place.

DIAPHRAGMATIC FLUTTER

Diaphragmatic flutter is a very rare condition of unknown etiology in which the diaphragm contracts rapidly, regularly and involuntarily. It resembles a tic such as is seen in other parts of the body. It

omes in paroxysmal form and may produce few, if any, symptoms On the other hand diaphragmatic flutter may sometimes be heard only when the patient is in a terminal state, as illustrated in Figure 555 The diagnosis can be made on fluoroscopic examination by detecting the rapid minute contractions superimposed on the normal excursions of the diaphragm It also gives rise to peculiar sounds that can be heard all over the chest. The possibility that the condition may be present will most often occur to the physician on hearing peculiar sounds during auscultation of the heart The sounds are usually low frequency and thus may be better appreciated using light pressure with the bell of the stethoscope The first time such a patient was observed by us it seemed that one was listening to two different hearts within the chest. In the case illustrated in Figure 556 there was evi

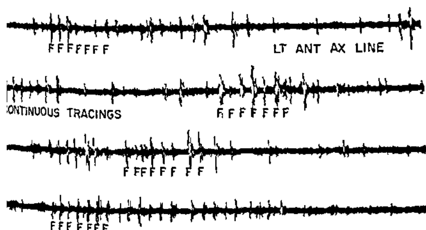
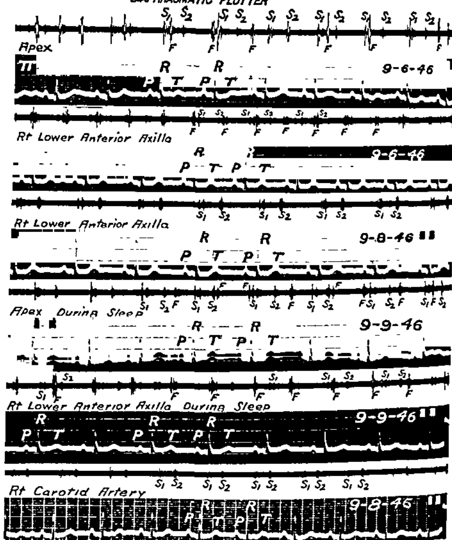


FIG. 555 Patient in a terminal state Had diaphragmatic flutter Flutter sounds (F) were clearly audible and louder than the normal heart sounds.

dence of rheumatic valvular disease However additional sounds were so prominent that before the true nature of the problem was determined the observers thought that atrial fibrillation was present. On more careful study it quickly became evident that fairly regular sounds entirely independent of the pulse were clearly audible in the right axilla, far removed from the heart Moreover, on auscultation over the carotid arteries the real heart sounds could be heard and counted at a normal rate of 75 It was then obvious that there were two origins of the various sounds heard over the chest one from the heart itself and the other from some other source possibly the diaphragm The latter mechanism was confirmed by fluoroscopic examination. Although these diaphragmatic contractions came and went without obvious cause they were present at times even during sleep The cause of this condition is still obscure



THORACIC DEFORMITY

Marked thoracic deformity such as is seen in kyphoscoliosis of the dorsal spine resulting from early postural defects, Pott's disease or poliomyelitis may be accompanied by slight systolic murmurs. So-called "funnel," "cup" or "saucer" shaped chests (*pectus excavatum*), however, frequently produce systolic murmurs (Fig. 557). The exact mechanism by which the sternal depression produces the murmur is debatable, but that a systolic murmur may be present over the mid-precordium without valvular disease or other common causes of murmurs is quite certain. Such cases generally ha ardiac disability,

hough when the deformity of the chest is marked a serious condition called cardiorespiratory failure may eventually develop Unfortunately, because of a systolic murmur and a posteroanterior chest film which indicates heart enlargement, such patients have been diagnosed as having organic heart disease. A lateral film of the chest reveals

d by the deformity and that its size

uer

uch as aneurysm of the

ciency)

y extensive pectus excavatum (funnel type) had a systolic murmur and, in addition, a grade

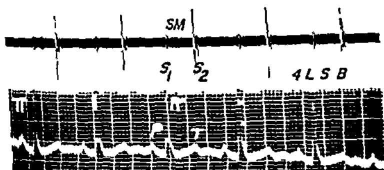


FIG. 557 Woman age 24 with a cup-shaped depression of the sternum. No history of rheumatic fever A grade III systolic murmur (SM) was heard best at fourth left interspace along outer edge of cup" This murmur was thought to be the result of chest deformity and of no clinical significance

III early, blowing diastolic murmur of aortic insufficiency that was heard best along the left sternal border He had spurious 'cardiomegaly' from the chest deformity and his palate was high arched but he exhibited no other features of Marfan's syndrome The presence of the diastolic murmur was a clue that his condition was a variant of the typical Marfan's syndrome with probable medial necrosis of the ascending aorta in the region of the aortic valve

MUSICAL SYSTOLIC MURMURS OF UNKNOWN ETIOLOGY

Victor McKusick s observations on muscial murmurs have greatly aided in our understanding of them however even after most exhaustive study the cause of some systolic murmurs remains unknown or obscure. This applies not only to murmurs of faint or moderate intensity but also to very loud ones The case illustrated in Figure 558 illustrates one of these puzzling problems This woman, 33 years old had primary pulmonary hypertension She had had occasional syncopal attacks for three years exertional dyspnea for two years

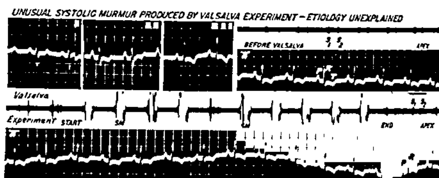


FIG. 558. Woman, age 33, with history of syncopal attacks, dyspnea and edema. Three standard electrocardiographic limb leads (upper tracing, left) show right axis deviation. Normal heart sounds (S_1, S_2) at apex (upper right). On Valsalva experiment (lower tracing) a loud, "leathery" grade VI murmur (SM) in late systole was induced, heard over entire precordium but maximum in the third and fourth left interspace. Note prompt disappearance of murmur (SM) with end of Valsalva experiment. Postmortem examination showed no congenital or acquired valvular heart disease, but right ventricular hypertrophy and extensive obliterative arteriosclerosis of the pulmonary arteries were present.

PECULIAR MUSICAL MURMUR OF UNDETERMINED ETIOLOGY (PERSISTING OVER 4 YEARS. UNCHANGED)

BEFORE EXERCISE

IMMEDIATELY FOLLOWING EXERCISE – CONTINUOUS TRACINGS SHOWING PRODUCTION AND DISAPPEARANCE OF MURMUR

FIG. 559 A 42 year old woman with rheumatic valvular disease. A very peculiar,

d dependent edema for six months On ordinary examination the art showed an accentuated pulmonary second sound and a very int apical systolic murmur The patient had noticed, at times peculiar sound within the chest. This could be reproduced at will by holding a deep breath, especially if she voluntarily compressed her chest at that time (Valsalva experiment) On auscultation an extremely loud grade VI somewhat musical systolic murmur could be heard all over the precordium loudest at the base of the heart. This would promptly disappear as the breath was released No adequate explanation could be offered Some months later she died,

LOUD MUSICAL MURMUR, TRANSIENT, OCCURRING IN LATE SYSTOLE RHEUMATIC MITRAL INSUFF & STENOSIS



Fig. 60 A 23 year old woman with rheumatic heart disease with mitral insufficiency and stenosis and atrial fibrillation. Had pansystolic apical murmur of mitral insufficiency but with some beats a peculiar loud musical late systolic murmur (SM) varying from grade I to VI was heard No relation to respiration or position Note transient loud late systolic murmur (SM) ending with second sound (S_2)

and on postmortem examination the heart itself showed no congenital or acquired abnormalities except an enlarged right ventricle There was generalized pulmonary arterial disease producing so-called chronic cor pulmonale Whether increased pressure in the pulmonary artery during the Valsalva experiment could produce vibrant murmur remains speculative

Another instance in which a loud musical systolic murmur has been observed to come and go without cause is seen in Figure 559 This patient was a woman 45 years of age with well compensated mitral stenosis and atrial fibrillation For several years she had been aware of a peculiar noise in her chest at times We have heard it at irregular

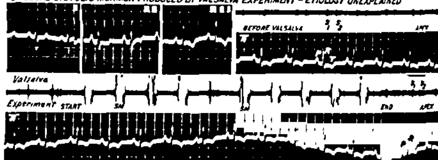


FIG. 558. Woman, age 33, with history of syncopal attacks, dyspnea and edema. Three standard electrocardiographic limb leads (upper tracing, left) show right axis

PECULIAR MUSICAL MURMUR OF UNDETERMINED ETIOLOGY (PERSISTING OVER 4 YEARS, UNCHANGED)

BEFORE EXERCISE

IMMEDIATELY FOLLOWING EXERCISE – CONTINUOUS TRACINGS SHOWING PRODUCTION AND DISAPPEARANCE OF MURMUR

FIG. 559. A 42 year old woman with rheumatic valvular disease. A very peculiar,

formed that at intervals a sound is produced during systole with similar to those of a stringed musical instrument. The situation may be analogous to that of a tuning fork which could be set in vibration or reinforced by sound waves of a certain frequency

SYSTOLIC "WHOOP" — NO HEART DISEASE

S1 S2 W P R T APEX CONTINUOUS TRACINGS

SAME PATIENT — SYSTOLIC CLICK

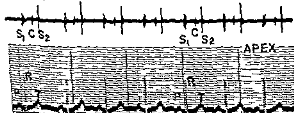


FIG. 567 Girl age 17 with no heart disease Had inconstant peculiar musical systolic sound (W) in the latter part of systole which resembled a "whoop" sound of whooping cough (upper tracing) The sound varied from grade V or VI to faint and sometimes was even absent. A systolic click (C) was frequently heard when the whoop sound was not present. It was not related to respiration or position.

and in phase with it At any rate these two cases appear to be similar both having the same etiology and both having definite mitral valve involvement with mitral insufficiency and mitral stenosis An other possible explanation for such a musical type of murmur might be that previous lung disease, such as pneumonia, pulmonary embolus

intervals, sometimes only for several seconds, at other times con-stantly, and then it might be absent for days. It was not related to position of the body or to breathing, and could always be brought out or increased both in its frequency and intensity by a brief effort. There was no x-ray evidence of diaphragmatic hernia or any other abnormality that might throw light on its causation.

A case somewhat similar to the one just described is shown in Figures 560 and 561. This was a 23 year old woman who had rheumatic heart disease with predominant mitral insufficiency associated

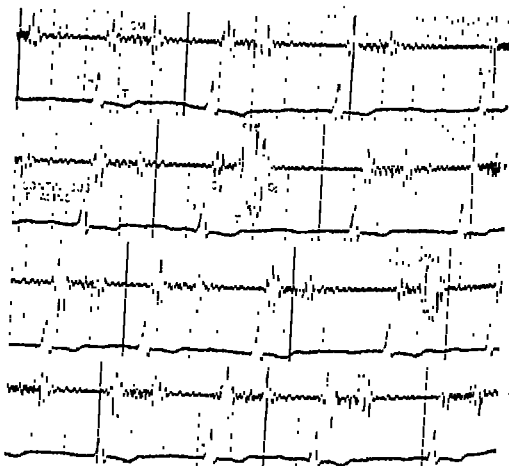


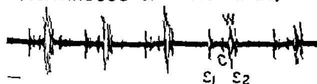
FIG. 561. Same patient as in Fig. 560: taken at fast speed. At times a late apical systolic murmur (SM) could easily be heard with the stethoscope off the chest

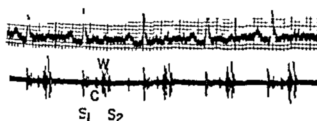
with mitral stenosis, atrial fibrillation and a giant left atrium. A transient, peculiar, loud musical systolic murmur was heard as an inconstant finding. A grade III to IV apical pansystolic murmur characteristic of mitral insufficiency was evident, but with some beats there was a peculiar, loud, musical systolic murmur that occurred in the latter half of systole, varying in intensity from grade I to grade VI. It was frequently grade V to VI, and it had no relation to respiration, but did seem to have some relation to effort.

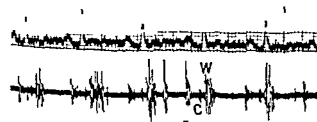
In the two cases just described one wonders about the possibility of mitral valve deformity with a cusp or chordae tendineae so de-

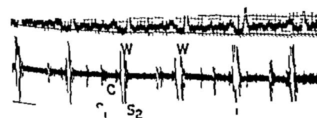
Systolic Whoop This is another form of a loud, inconstant musical murmur that occurs in the latter part of systole in patients without any apparent or significant heart disease The murmur is similar to the "whoop" of whooping cough This sound was illustrated by a

VARIATION OF SYSTOLIC "WHOOP"
(CONTINUOUS TRACING-APEX)



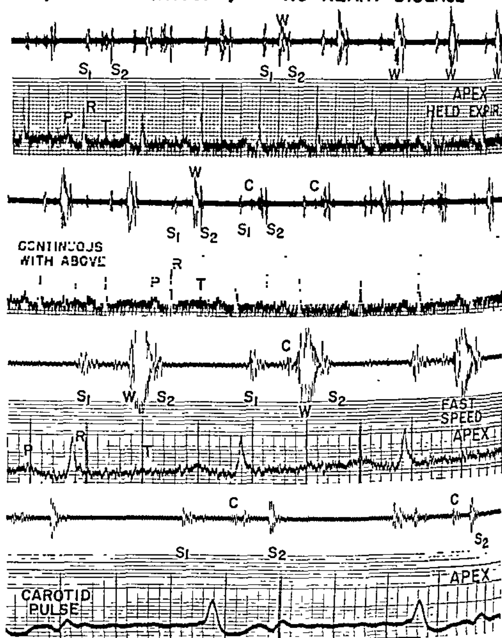








intensity of systolic
(C) Respiration is



pleurisy, etc., produced adhesions in an area adjacent to the ventricles of the heart. At times air might be trapped in areas of the lung adjacent to the heart. Then, under certain circumstances, ventricular contractions may produce an impact on the lungs that forces air out through a bronchus, causing these murmurs. At any rate, this loud, late musical murmur is usually of no serious consequence. The prognosis in the individual case would depend upon that of the underlying heart disease and not the presence of this type of murmur.

... h no heart disease who was evaluated for pos-

At times she had no sounds other than her two normal heart sounds

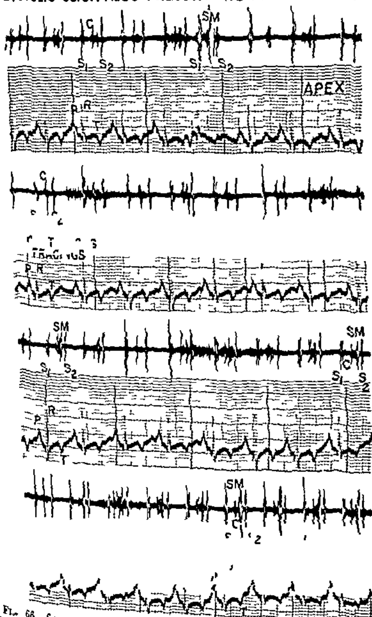


FIG. 68 6 year old girl with an inconstant late apical musical systolic murmur (SM). A systolic click (C) occurring between first and second sounds (S_1 S_2) was usually present, but occasionally absent.

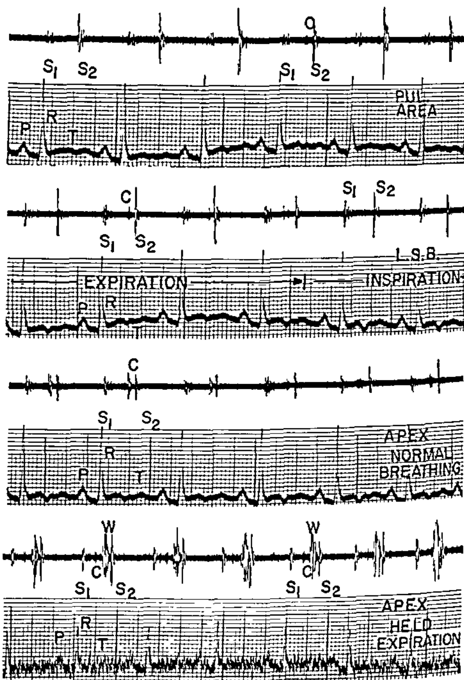


Fig. 565. Same patient as Fig. 563. Late musical systolic murmur (systolic

tion. It was generally heard best with the patient sitting. It was repeatedly demonstrated, however, that when the patient was sitting upright firm pressure with the hand over the left precordium could cause this peculiar musical murmur to disappear. The murmur was

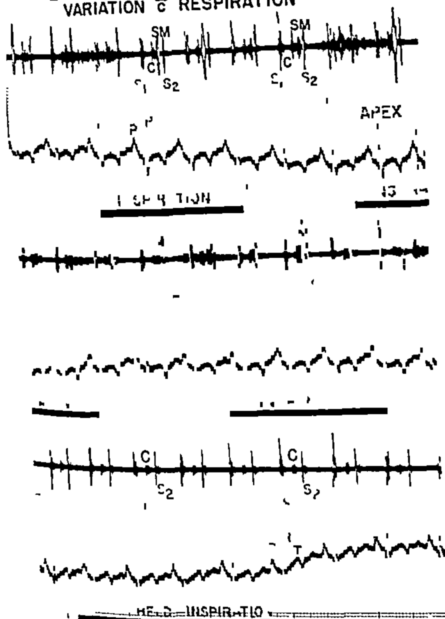
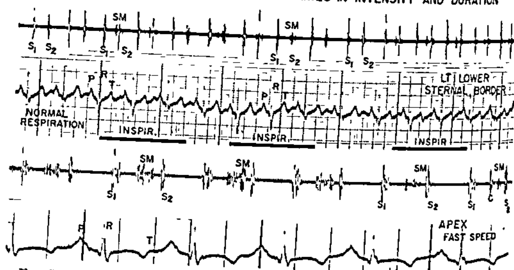


FIG. 568. Same patient as Fig 566 showing variation of late musical systolic murmur (SM) and systolic click (C) with normal respiration. With held inspiration (lower strip) musical murmur absent, but systolic click (C) remained

at other times a systolic clicking sound was heard. At times a ver loud (up to grade V), apical systolic musical sound simulating "whoop" of whooping cough was present (Fig. 562). This seemed t bear no relation to respiration or the position of the patient. This finding was considered benign, and the patient was reassured.

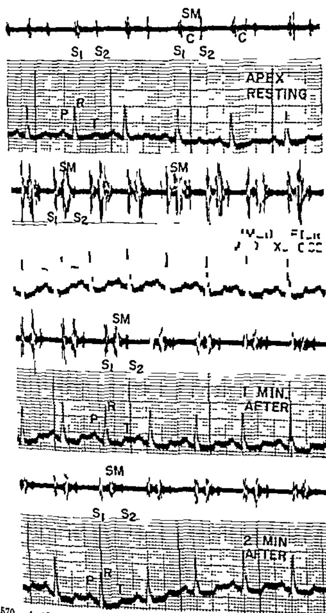
A similar patient (Figs. 563, 564, 565) was a 30 year old physician who had been observed over a five year period. He was first examined when a medical student, and findings similar to those in the patient just described were present. His "whoop" sound was not as loud, but would attain grade III to IV intensity. A systolic clicking sound was heard frequently. As in the previous case, there was no evidence of heart disease. Since we first described this condition, other physicians have told us of similar cases. One wonders whether a congenital



... click (C).

anomalous band may be present in the ventricle near the outflow tract. Such a band at times could vibrate in a manner similar to a musical instrument, as previously discussed.

Still another example of a late musical murmur was heard in a six year old girl who also had a sound in midsystole. Her musical murmur appeared to be related to inspiration and body position. An x-ray examination of the heart was normal. The electrocardiogram was thought to be normal except for some T wave variations which could be altered by normal respiration. As shown in Figures 566, 567 and 568, coincident with inspiration the systolic sound appeared to become more prominent, attaining grade IV in intensity. This sound likewise had a musical quality and was in the latter part of systole. With a deep held breath the murmur disappeared but the midsystolic sound remained. In addition, the murmur could be altered by change in posi-



G. 570 A 46 year old firem n ... ant musical systolic murmur ... systolic click (C) was noted ... y after exercise a transient ... ually waned in intensity after

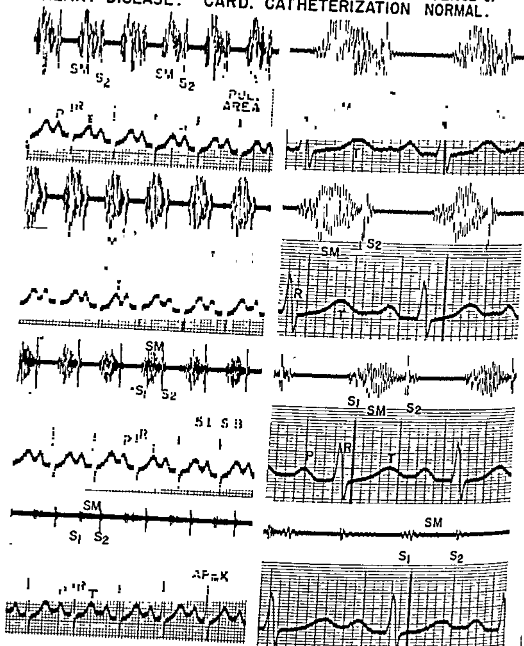


FIG. 569. A 13 year old girl with a known murmur since early childhood. Had grade IV musical systolic murmur (SM) loudest at third left sternal space Left column tracings at normal speed; right column, same area at fast speed Note loud systolic murmur (SM) over pulmonic area and left sternal border; heard faintly at apex. Second sound (S_2) normally split over pulmonic area (upper tracing). Patient was asymptomatic.

interpreted as most likely of extracardiac origin, since it could be altered by external pressure and position. Possibly it was produced by the systolic impulse of the heart temporarily forcing trapped air from adjacent lung tissue through a bronchus. The possibility of a congenital anomalous band or anomalous chordae tendineae cannot be ruled out. This and the entire group of late musical systolic murmurs

it was felt that this murmur was benign, and the girl was told to lead a normal life.

Another example of a transient, late, apical musical systolic murmur was heard in a 46 year old fireman who was referred for evaluation of possible congenital heart disease At times a loud systolic murmur had been heard, and at other times there was no murmur whatsoever The patient was found to have coronary artery disease with angina pectoris A double Master tolerance test was definitely positive In addition he was noted to have a transient late musical systolic murmur which could usually be precipitated by effort (such as an exercise tolerance test or climbing four to five flights of stairs) While resting only a slight systolic murmur, grade I to II was evident (Fig 570) In addition, a systolic sound (or click) was present Immediately after moderate exercise a late, grade IV musical murmur was heard which occupied the latter half of systole It was most marked in the middle of systole and tapered off by the end of systole After total cardiovascular evaluation it was decided that the patient had arteriosclerotic heart disease with angina pectoris and that the peculiar musical systolic murmur had no relation to this diagnosis Again the possibilities of a congenital anomalous band arose He was reassured concerning the benignity of the musical murmur, and treatment was aimed at his underlying basic coronary artery disease

edical advice. He had a recent history of trauma (automobile accident), and later had a myocardial infarction A systolic murmur over the precordium and right neck suggested the possibility of aortic stenosis. However in addition, a loud rough, musical, somewhat late systolic murmur was frequently but not constantly heard It could be made to disappear by deep held inspiration of a Valsalva maneuver

THE DURATION OF VENTRICULAR SYSTOLE

The duration of mechanical ventricular systole can be measured fairly well by the length of the interval between the first and second heart sounds The length of electrical systole is measured by the Q–T interval in the electrocardiogram Although the durations of mechanical and electrical systole in most cases are almost the same and vary in a parallel fashion this is not always true It was hoped that one could predict the length of the Q–T interval by sensing the duration of the interval between the first and second heart sounds (systole) as compared to that between the second and first sound (diastole) In fact occasionally we have been able to predict by auscultation that this interval [illegible] d (Fig 572) Such cases of uremia or of the blood is low

have been regarded as benign. They are frequently associated with a systolic sound or click. The murmur has been inconstant, musical and late in systole. In contrast, the murmur of organic mitral insufficiency has been characteristically constant and pansystolic in time, beginning in early systole.

Such musical murmurs are often misinterpreted as due to a congenital heart lesion, such as ventricular septal defect. This diagnosis was entertained in the case of a 13 year old girl who had had a known murmur since early childhood (Fig. 569). A grade IV systolic mur-

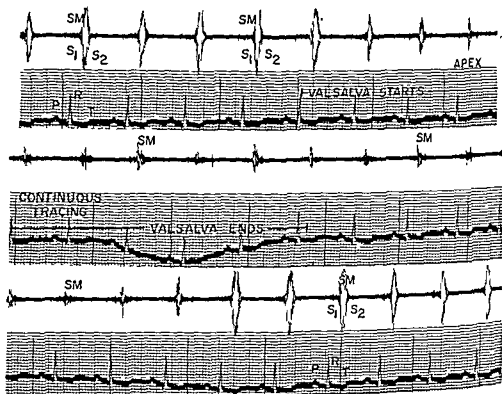
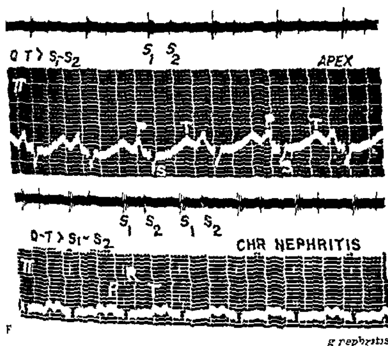


FIG. 571. Man, age 64, with history of myocardial infarction Had systolic murmur (SM) grade III, suggesting aortic stenosis, heard over precordium. A superimposed, inconstant, loud, late, musical systolic murmur (SM) was noted, which disappeared with deep-held inspiration or with Valsalva maneuver.

mur was heard over the precordium, best at the third left sternal space. A palpable thrill was likewise present. The second heart sound was split and of normal intensity. The degree of splitting increased with inspiration in normal fashion. Electrocardiogram was normal, and x-ray showed some fullness in the region of the pulmonary artery segment. The murmur did have a somewhat musical, rough quality, and the diagnosis of ventricular septal defect was entertained. Cardiac catheterization, however, revealed no evidence of shunt between the ventricles. The final impression was that this murmur, although congenital, was not due to a septal defect or pulmonic stenosis, but was rather of unusual etiology and possibly due to an anomalous band.

heart sounds very distant where they ordinarily should have been well heard, but very loud borborygmi were frequently heard in the left anterior thorax (Fig 575) In fact this combination of auscultatory findings immediately suggested the diagnosis in a recent case Had not attention been paid to these simple auscultatory points, the diagnosis would have been overlooked When the noise of rumbling gas in the stomach or intestines is prominent, it temporarily obscures the heart sounds.



F g nephritis (Ca)

Note mechanical an electrical systole.

ARTERIOVENOUS FISTULA

Arteriovenous fistula produces a characteristic continuous murmur with both systolic and diastolic components Frequently the systolic phase is the louder but this is not always the case Patent ductus arteriosus is truly an arteriovenous fistula but any acquired communication between large arteries and veins can cause a similar murmur Traumata such as gunshot or stab wounds are common causes of this condition though there are infrequent acquired non

(Fig. 572). However, the lack of parallel between mechanical and electrical systole is well shown in Figure 573. In one of the cases the interval between the first and second sound was quite short (0.24 to 0.28 second) while the Q–T interval was very long (0.48 second). In another case the reverse was true. It follows that auscultation may be, but is not always, an aid in estimating the duration of electrical sys-

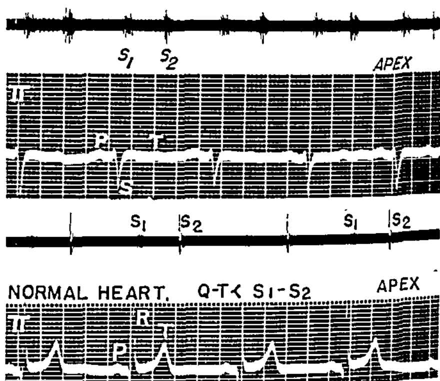


FIG. 572. Upper tracing: Woman, 45 years old, with chronic nephritis. (Blood calcium = 2.2 mEq.) Slightly prolonged Q–T interval suspected on auscultating long mechanical systole (S_1, S_2). Lower tracing: Woman, age 22, with no heart disease. On auscultation the interval between first (S_1) and second (S_2) sounds seemed somewhat prolonged. Note lack of correlation between mechanical and electrical systole, the latter being shorter.

tole, though it does serve as a fair guide in predicting the duration of mechanical systole.

DISPLACEMENT OF THE HEART TO THE RIGHT

If the heart is markedly displaced into the right thorax, or in cases of dextrocardia or mesocardia, it is obvious that the heart sounds will be less prominent over the left than the right anterior thoracic wall. In the case illustrated in Figure 574, the heart sounds were almost inaudible near the left nipple but were quite loud on the right side. On x-ray study eventration of the left diaphragm was found, the stomach lying in the left lower thorax while the patient was supine. One might have suspected this condition because not only were the

traumatic instances Figure 576 illustrates a continuous murmur obtained over the left antecubital region of a young man of 30 As a boy, he fell and injured his left arm In the course of time the arm became a little larger and warmer, and extensive arteriovenous com m cations developed around the left elbow A loud continuous murmur was audible in this region Congenital coronary–arteriovenous fistula over the left ventricle producing a similar murmur has already been discussed

There are other conditions in which murmurs are heard in the peripheral part of the body, and the question of arteriovenous com



gnosis.

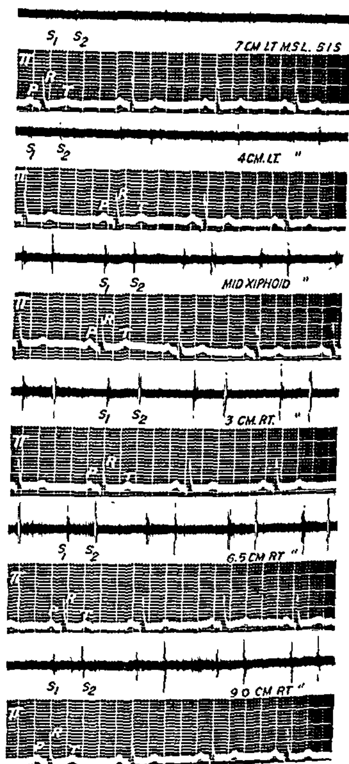




FIG. 5 6 Man age 30 who had three years of active service in the Army Signal Corps. Had grade V continuous murmur over a v fistula of left antecubital region.

munication arises If the murmur is continuous the evidence points strongly to one or more fistulous tracts between arteries and veins However if the murmur is not continuous but coming with each systole, one would be more tempted to regard the condition as an aneurysm or a hemangioma In the case illustrated by Figure 577, a prominent late systolic murmur was heard over the right temporal bone There was no murmur on the left and there were no significant murmurs over the heart The diagnosis here has not been confirmed but the possibilities both of a hemangioma and a cranial arteriovenous fistula still need to be considered

Sometimes the presence of an unexplained murmur in other parts


HEART DISPLACED INTO RT. CHEST – SOUNDS BEST HEARD TO RT OF STERNUM
S1 S2
7 CM LT MSL. 5 I S
II
R
P
T
S1 S2
4 CM. LT. "
MID XIPHOID "
3 CM. RT. "
6.5 CM RT "
9 0 CM RT "

CONTINUOUS MURMUR OVER LEFT EYEBALL

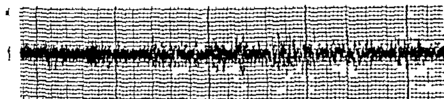
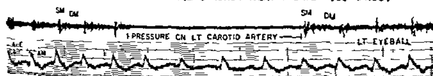


FIG. 578 Young girl with continuous murmur heard over left eyeball Angiogram showed a vascular malformation in the left temporal area



ery

ONTINUOUS MURMUR OF A-V FISTULA BETWEEN
LT CAROTID ARTERY & CAVERNOUS SINUS

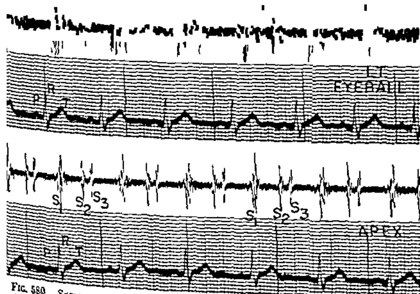


FIG. 580 Same patient as in Fig 579 Continuous murmur (SM-DM) over left eyeball (also over left maxillary region) Had constant third sound (S_3) at apex (lower tracing) which had a gallop quality

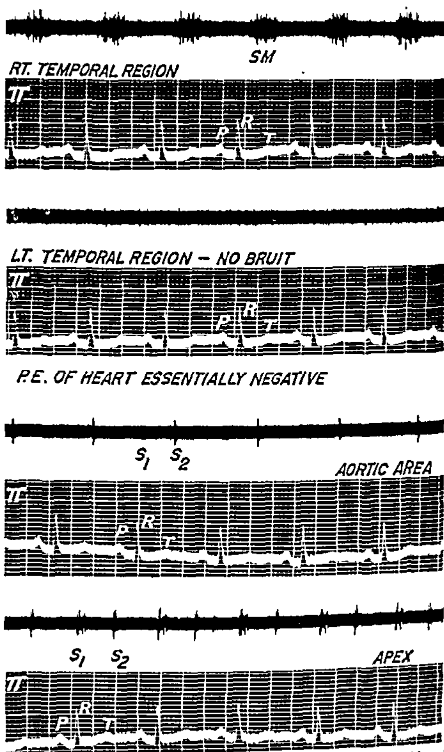
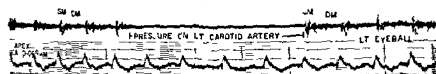


FIG. 577. A 45 year old woman with trigeminal neuralgia and probable arteriovenous aneurysm in right temporal region. Four years previously she had an operation for division of fifth cranial nerve. A definite grade II high pitched systolic bruit (SM) was present over the right (upper tracing) but absent over the left temporal region (second tracing). Heart was not remarkable (lower two tracings)

CONTINUOUS MURMUR OVER LEFT EYEBALL



Fig 578. Young girl with continuous murmur heard over left eyeball Angiogram showed a vascular malformation in the left temporal area



artery

CONTINUOUS MURMUR OF A-V FISTULA BETWEEN LT CAROTID ARTERY & CAVERNOUS SINUS



Fig. 580 Same patient as in Fig 579 Continuous murmur (SM–DM) over left eyeball (also over left maxillary region) Had constant third sound (S_3) at apex (lower tracing) which had a gallop quality

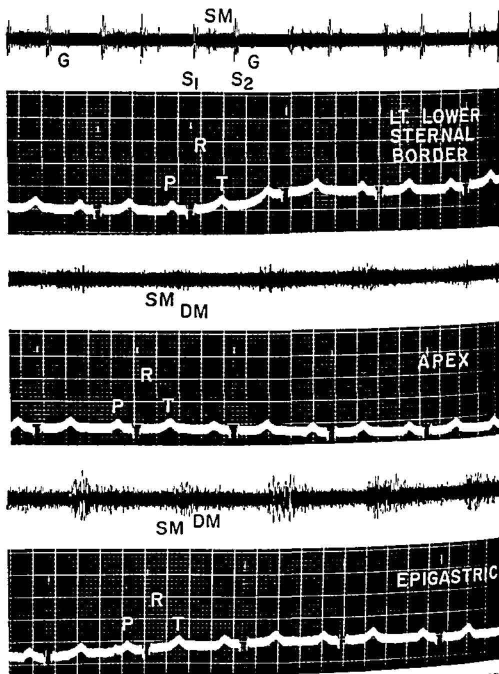
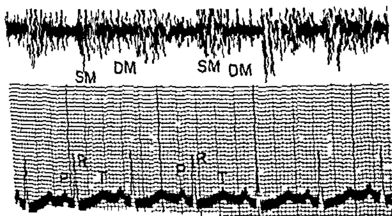


FIG. 58
over the
tracing);
(SM) and ventricular diastolic gallop (G) were present. Had early congestive fail-
resulting from traumatic arteriovenous fistula (gunshot wound of the abdomen).
Following operation the continuous murmur disappeared, as did all signs of failure

of the body serves as a diagnostic clue Such was the case illustrated in Figure 578 A continuous murmur was heard over the left eyeball in a nine year old girl Angiograms showed a vascular malformation in the left temporal area After this experience routine auscultation over the eyeballs was carried out on normal subjects, and it was discovered that systolic as well as continuous murmurs may be present in the absence of any known brain lesions However, a loud murmur heard over the eyeball or skull should suggest some diseased state This is well shown by the case of a 12 year old boy illustrated in Figur -79 nd 580 A continuous machinery murmur, grade IV, was heard on and over the left eye ball to this area a congenital



arteriovenous fistula between the left carotid artery and cavernous sinus was diagnosed Pressure on the right carotid artery caused the murmur to diminish or disappear (Fig 579) The patient's left eye ball protruded and auscultation of the heart at the apex revealed a constant third sound that had the quality of a ventricular gallop (Fig 580) This sign brought up the possibility of early cardiac decompensation It is urged that the stethoscope be used over areas other than the chest A 25 year old man was seen on a routine Army Reserve (National Guard) physical examination (Fig 581) The examining physician noted a faint continuous murmur over the apex and lower sternal border On auscultating over the epigastrium a moderately loud continuous murmur was heard Early congestive heart failure was present as evidenced by a ventricular diastolic gallop pulsus alternans and cardiomegaly A faint systolic murmur

HEMANGIOMA RIGHT CALF C̄ BRUIT (NOT CONTINUOUS, ON AUSCULTATION)

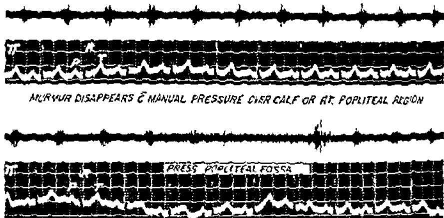
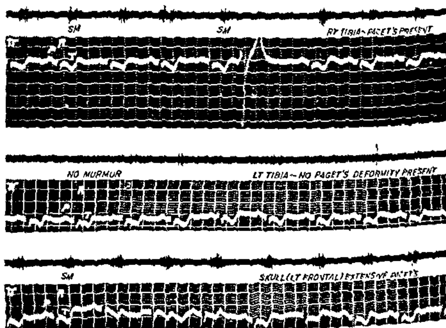


FIG. 583. A 29 year old woman who had noticed a "lump" in her right calf for five years. She had a *grade III high* pitched, noncontinuous murmur over right

PAGET'S DISEASE OF SKULL AND RIGHT TIBIA C̄ PRODUCTION OF MURMUR OVER THESE AREAS



was heard at the apex and the second sound over the pulmonary area was accentuated. Reference to the patient's history revealed a gunshot wound of the abdomen. A diagnosis of arteriovenous fistula was made and confirmed at operation, where the renal artery was found to be involved. Following the operation the murmur disappeared, as did all signs of heart failure.

An arteriovenous fistula that had been purposely created by surgery a number of years previously was discovered on routine physical examination of a 15 year old patient with mental retardation (Fig 582) The murmur associated with a palpable thrill, was loud, grade IV to V and heard best over the right neck Apparently the operation had been performed in an unsuccessful effort to improve blood flow to the brain of this mentally deficient child

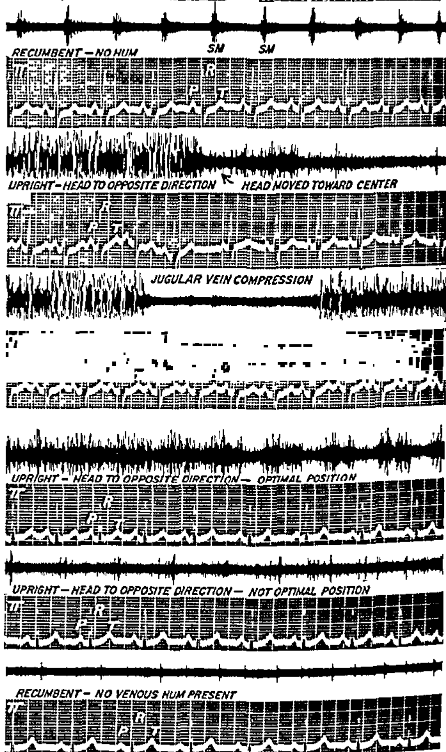
The patient illustrated in Figure 583 a 29 year old woman, is one in whom the diagnosis of hemangioma was proved by operation In this case the surgeons first thought they were dealing with an arteriovenous fistula A loud murmur was audible over the right calf but was not continuous, being limited to late systole The absence of the classic murmur of the arteriovenous fistula might have led one to doubt the accuracy of the surgeon's diagnosis It is of interest that only moderate pressure with the stethoscope over the area was needed to obliterate the murmur This is in accord with the pathologic finding of an extensive capillary bed in the tumor

It has recently been suggested that bone lesions in active Paget s disease resemble minute arteriovenous fistulae With this in mind a patient with a classic case of advanced Paget s disease was examined with care and a systolic murmur was found over the skull and right tibia (Fig 584) It is significant that no murmur was heard over the uninvolved left tibia In this case the murmur in the right tibia appeared to be brought out by walking for it was not detected during a previous rest period

VENOUS HUM

A continuous humming murmur in the neck is a common finding in children but less so in adults It has no pathologic significance The sound is heard best in the lateral areas of the supraclavicular fossae (Fig 585), and its origin has been regarded as the jugular vein It is more readily heard when the patient is upright rather than recumbent and when his head is turned away from the examiner As a rule the sound is heard better in the right supraclavicular fossa During auscultation, the right hand should hold the bell of the stethoscope over the right supraclavicular area while the left hand grasps the patient s chin from behind and tilts it upward and to the left At an optimal position when a ' stretch has been produced in the region of the jugular vein on the right, the hum will be heard It often has the character of a continuous roaring sound and it can be made to disappear by moving the head forward again Light pressure with the finger over the upper part of the jugular vein also will cause the continuous murmur to disappear

The venous hum probably results from rapid downward flow through the artificially stenosed jugular vein Although continuous, the murmur often is loudest in early diastole It may envelop the



pressure on . . . . Woman, age ... with no hear . Note loud continuous macl ... is turned to opposite direction. Second tracing: Hum less prominent; head turned near to midline. Lowest tracing: Venous hum absent with patient lying flat and head forward.

second sound, as in patent ductus arteriosus In children the hum is often loud and may be heard well even if the child is sitting with his head forward Characteristically, however, a recumbent position causes the disappearance or marked diminution of the murmur It may be transmitted over the upper precordial area It is easy to see how the murmur would then be confused with that of patent ductus (Figs 586 587) In fact, we have observed cases referred for surgery for patent ductus because of this transmitted venous hum In such cases the correct diagnosis would readily become clear if pressure over the jugular vein caused disappearance of the continuous murmur A change in position of the head might likewise cause the disappearance (Fig 587) or reappearance of the continuous murmur, which would not be the case in patent ductus Also, in these cases

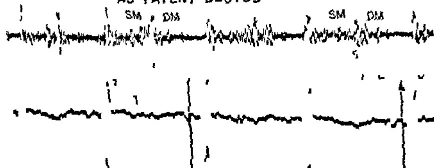


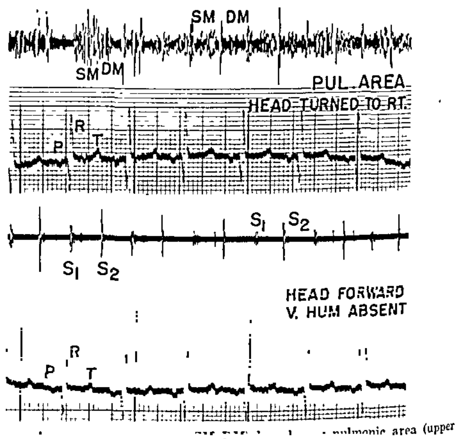
FIG. 586. A 2½ year old boy with a grade III to IV continuous murmur loudest over the pulmonic area. Note continuous murmur (SM–DM) enveloping the second sound (S_2) Pressure over the jugular vein caused this murmur to disappear

auscultation over the supraclavicular fossa will generally reveal that the continuous murmur is loudest over this area

A venous hum is common in conditions associated with a rapid circulation and a high cardiac output It is present with anemia, beriberi heart disease, thyrotoxicosis and during pregnancy (particularly during the last two trimesters) Figure 588 represents two patients who had a venous hum during pregnancy

One patient was referred to our cardiac clinic because of a continuous murmur over the first and second left interspaces This was before the era of cardiac catheterization and the patient had been to numerous clinics Various diagnoses such as arteriovenous fistula pulmonary a v fistula and patent ductus arteriosus had been entertained Light pressure over the left jugular vein completely eliminated the murmur thereby establishing the diagnosis as venous hum Unfortunately this patient had been treated for a number of years as having some type of congenital vascular anomaly and a deep set cardiac neurosis was already present.

Of particular importance is the finding of a venous hum with thyrotoxicosis. Over the past ten years we have made a careful search for venous hum in all of our patients with thyrotoxicosis and have noted the hum in most of them. As a rule the sound has been heard best in the right supraclavicular fossa using the technique described above. The *absence* of venous hum in a patient suspected of having thyrotoxicosis has, in our experience, made unlikely the possibility of



... pulmonic area (upper

thyrotoxicosis. On the other hand, venous hum may frequently be present *in normal adult patients without thyrotoxicosis* or other evidence of disease. These observations have often been verified by laboratory studies, including protein-bound iodine and radioactive iodine uptake.

Figure 589 shows a composite of five patients with thyrotoxicosis and venous hum. It will be noted that the first three patients and also the last one show a diastolic accentuation of the hum, whereas

in the fourth the murmur enveloped the second sound in a manner more typical of patent ductus arteriosus In fact, our observations of patients with thyrotoxicosis reveal that the so-called thyroid bruit is often actually a venous hum Some patients though will have a bruit that consists of a systolic murmur in the region of the thyroid which is felt to be an accentuation of the normal flow through the arteries in that area This becomes clear when the venous hum is

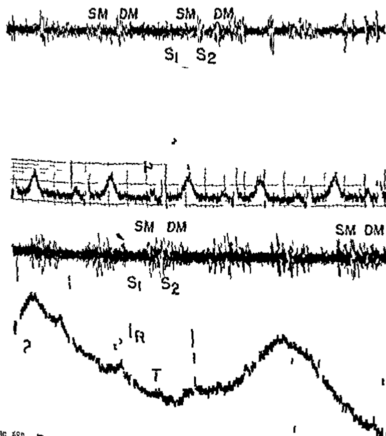


Fig 588 Two women (upper and lower strips) with prominent venous hum associated with pregnancy Note continuous murmur (SM–DM) in each case

eliminated by pressure and the systolic murmur over the carotids persists (Fig 590) Less commonly, an actual continuous murmur

bruit in thyrotoxicosis The hum at times may be quite loud—in fact a palpable thrill may be felt over the area However the patient is

unaware of its presence and actually does not hear it. One patient, a 36 year old woman, was seen whose chief complaint was a roaring noise in her head. The only significant finding was a loud venous

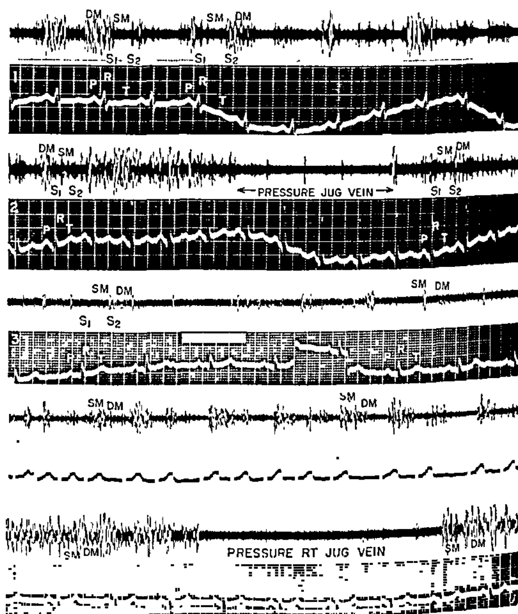


FIG. 589 Composite of five patients with thyrotoxicosis and a prominent venous hum. Note continuous murmur (SM–DM) in each case. Jugular vein compression causes disappearance or decrease in intensity, as illustrated in patients in second, third and fifth tracings.

hum which was unquestionably causing her symptoms. When the examiner listened to the hum with the stethoscope and caused it to disappear by jugular compression, the patient could state exactly when the "noise in her head" disappeared or returned. The noise was

THYROTOXICOSIS — AUSCULTATORY FINDINGS IN NECK

VENOUS HUM

RT SUPRCLAV FOSSA

PRESS RT JUG VEIN

SM

S1 E S2

SM

S1 E S2

RT CAROTID ART

SM

S1 S2

SM

E

S1 S2

LT CAROTID ART

VENOUS HUM AUDIBLE TO PATIENT AND CAUSING ANNOYANCE

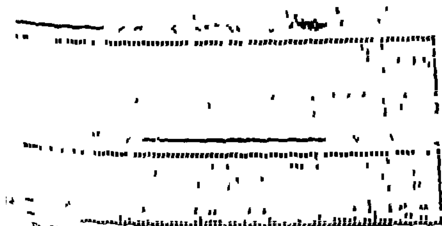


Fig. 591 [illegible] 56 year old woman with loud venous hum that was audible and annoying to h[illegible] Note disappearance of hum with jugular vein compression as shown on continuous tracing Patient could immediately *tell* when hum disappeared coincident with jugular vein compression.

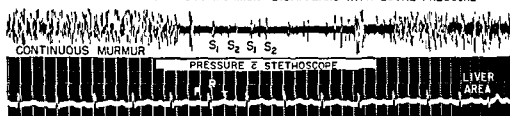


FIG. 592. Patient with cirrhosis of the liver and continuous murmur heard in right upper quadrant. Pressure with the stethoscope caused disappearance of the continuous murmur.

CIRRHOSIS OF LIVER c̄ CONTINUOUS MURMUR



CONTINUOUS MURMUR

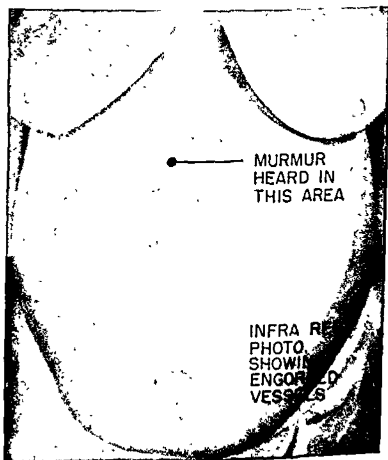


FIG. 593. Woman with cirrhosis of the liver A continuous murmur was heard in the area shown.

so great that it was a source of great bother and worry to the patient Figure 591 illustrates both the presence of this loud venous hum and its disappearance with jugular vein compression A "choker" fashioned out of light plastic was fitted around her neck to compress the region of the jugular vein The patient wore this "choker" and it relieved her complaint The alternative treatment if this had not been successful would have been actual ligation of the vein

Venous Hum over the Liver In patients with cirrhosis of the liver a continuous murmur is sometimes heard over the liver or over an adjacent area in the epigastrium Two patients with Cruveilhier-Baumgarten syndrome who demonstrated this sound are shown in Figures 592 and 593 In the first patient (Fig 592) a prominent continuous murmur (venous hum) was heard over the liver Pressure with the stethoscope caused its immediate disappearance, and the heart sounds were then well heard On release of pressure the continuous murmur returned This phenomenon is similar to the venous

sociated with cirrhosis

CONTINUOUS MURMUR OVER THE LACTATING BREAST

A continuous murmur may be heard over the breast of a pregnant woman or more commonly one who is nursing her child (Fig 594)

CONTINUOUS MURMUR OVER LACTATING BREAST

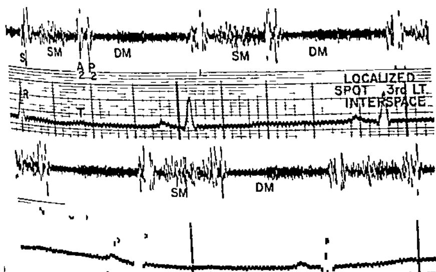


FIG. 594 A 24 year old woman with a continuous murmur heard over a lactating breast (SM DM) Peak of systolic murmur is in middle rather than late systole and continuous murmur does not envelop second sound Diastolic component (DM) is louder in second half of diastole.

This is of a continuous quality and can be made to disappear b pressure with the finger or stethoscope over the area. It is often hea only over one breast. The sound has been confused with patent duct arteriosus, pulmonary arteriovenous fistula and other forms of co genital heart disease having continuous murmurs. Patent ductus mostly likely to be erroneously diagnosed when the murmur is ove the left breast. We have assumed the mechanism to be similar to tha of a venous hum in the neck, and believe that the flow through th engorged veins of the breast produces the continuous murmur. Th exact etiology of the murmur is somewhat in question, and som observers have felt that it may be arteriovenous rather than strictl venous. At any rate, it is benign, and the patient can be reassured. Its importance lies in its possible confusion with the murmurs of patent ductus or arteriovenous fistula.

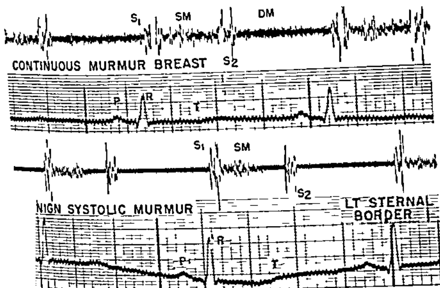
AUSCULTATORY FINDINGS IN PREGNANCY

As mentioned earlier, in our discussion of systolic murmurs, a functional systolic murmur is a frequent finding in pregnant patients. These sounds, ranging from grade I to III, are a common occurrence and sometimes are misinterpreted as being due to organic heart disease. They are most likely to be heard during the last trimesters of pregnancy, when they can be auscultated along the left sternal border, at the apex, or over the base of the heart. A diastolic murmur, unlike these benign systolic sounds, would indicate underlying organic heart disease; but a systolic murmur of grade III or less is best classified as functional during pregnancy, unless, of course, there is other evidence of heart disease (Fig. 595). As previously discussed, during pregnancy a venous hum may be heard over the neck, and, after lactation has begun, a continuous murmur over the breast is a frequent finding.

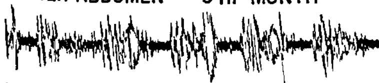
A normal physiologic third heart sound is more evident during the course of pregnancy. Also, in some patients, extrasystoles will appear for the first time during pregnancy. This latter finding may be disturbing to the patient, and organic heart disease may be wrongly suspected, particularly when, in addition to the extrasystoles, there is a systolic murmur and a heart that appears to be slightly larger than normal because of the elevation of the diaphragm with pregnancy.

Maternal Souffle. This sound, a continuous murmur that is usually high pitched and musical, is illustrated in Figure 596. It is a constant finding in pregnant patients and should present no problems in differential diagnosis. A systolic murmur of the type commonly heard over the large arteries is also a characteristic finding over the lower abdomen of the pregnant patient, particularly during the latter trimesters.

SOME AUSCULTATORY FINDINGS OF PREGNANCY



MATERNAL SOUFFLE HEARD OVER LOWER ABDOMEN - 9th MONTH



G. 96 Woman age 25 illustrating maternal souffle heard over lower abdomen (Phonocardiogram from tape recording)

FETAL HEART SOUNDS

The detection of fetal heart sounds is a common practice in both he diagnosis of pregnancy and the determination of the viability of he fetus. There are even rare instances in which diagnosis of heart

This is of a continuous quality and can be made to disappear pressure with the finger or stethoscope over the area. It is often hea only over one breast. The sound has been confused with patent duct arteriosus, pulmonary arteriovenous fistula and other forms of co genital heart disease having continuous murmurs. Patent ductus mostly likely to be erroneously diagnosed when the murmur is ov the left breast. We have assumed the mechanism to be similar to tha of a venous hum in the neck, and believe that the flow through th engorged veins of the breast produces the continuous murmur. Th exact etiology of the murmur is somewhat in question, and som observers have felt that it may be arteriovenous rather than strictl venous. At any rate, it is benign, and the patient can be reassured. It importance lies in its possible confusion with the murmurs of paten ductus or arteriovenous fistula.

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There is reason to suspect that there is a fundamental misconception when adult heart sounds are described as resembling 'fetal rhythm' By this phrase it is inferred that the two sounds are alike and that systole and diastole are equal in length On ordinary auscultation only one fetal heart sound is heard and the sounds obviously will all be alike. In the case illustrated in Figure 597 one of the sounds was almost inaudible on ordinary auscultation In other words when

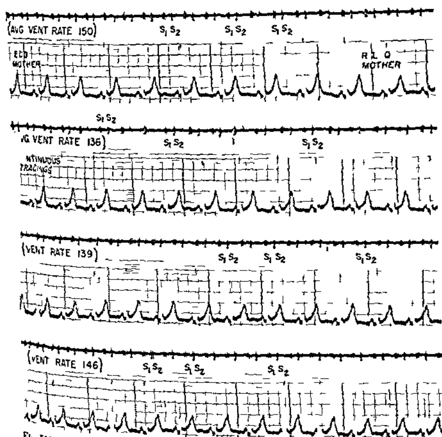


Fig. 599 Same patient as Fig 598 *Continuous tracings of fetal sounds showing sinus arrhythmia one week before birth of normal healthy girl*

adults manifest a so-called 'fetal rhythm' the first and second sounds are alike, whereas in the fetus the two sounds differ greatly in intensity Nevertheless, the phonocardiograph may be useful in detecting fetal sounds that otherwise are inaudible

AORTIC ANEURYSM

The auscultatory signs of aneurysm of the aorta vary depending on the site of involvement and the pathologic changes inherent in the

block has been made prenatally on finding a constant fetal bradycardia. Fetal heart sounds are generally not audible until about the fifth month. An example of such sounds in a woman six months pregnant is shown in Figure 597. These were heard over the lower midabdomen. In this instance it was not clear which was the first and which the second heart sound, although it is likely that the loud sound

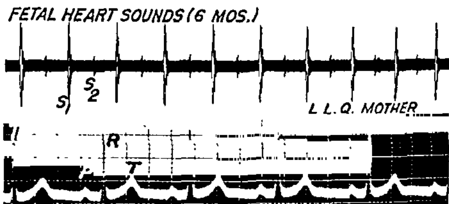


FIG. 597. Fetal heart sounds in a 32 year old woman, six months pregnant. Electrocardiogram of mother; heart sounds of fetus Note presence of two distinct sounds (S_1, S_2) with marked difference in intensity. This was clearly audible clinically.

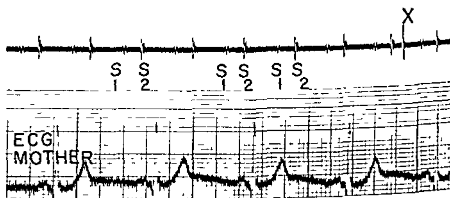
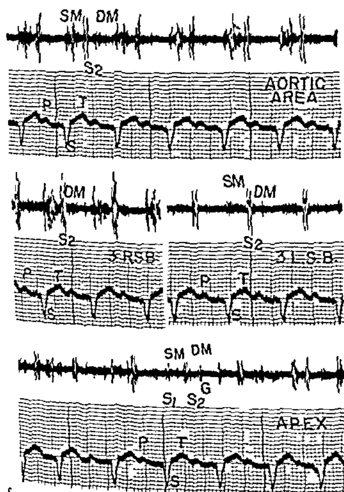


FIG. 598. Fetal heart sounds (S_1, S_2) of baby girl one week prior to birth. Note second sound (S_2) is louder than first (S_1). (X) is artefact due to fetal movement.

was the second. In Figure 598 the distinct fetal heart sounds one week before birth are shown, and here the second sound is the louder. In Figure 599, the fetal heart beat three days before birth was observed clinically to have sinus arrhythmia, varying in rate as much as 20 beats a minute. This afforded an opportunity to determine the first heart sound by means of the change in the length of diastole. In this case the second sound was the louder. At birth a healthy baby girl showed no evidence of cardiac anomaly.

particular aneurysm Aneurysm involving the ascending aorta, but without aortic valve involvement, may be associated with no ausculta tory signs, particularly if the aneurysmal dilatation is minimal or moderate With more significant involvement a systolic murmur is frequent over the aortic area, and the aortic second heart sound may be accentuated (Figs 600–602) Sometimes it has the so-called tambour quality which used to be regarded as a sign of syphilitic aortitis. However, we have found this sound in patients with hyperten

LUETIC ANEURYSM ASC AORTA c̄
"RT SIDED" AORTIC DIAST MURMUR



...ansmitted diastolic murmur (DM) and diastolic gallop (G)

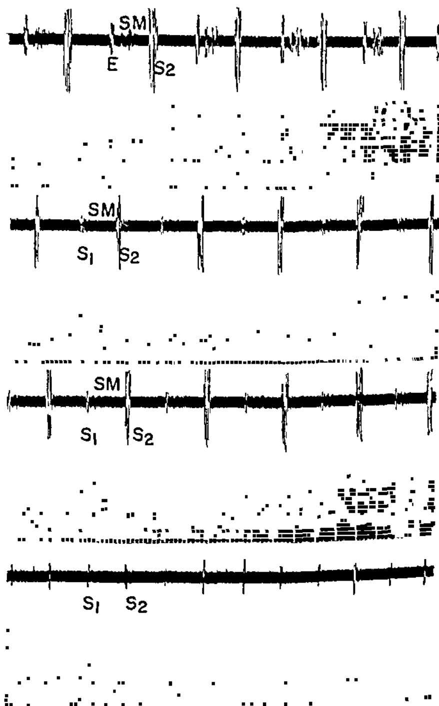


FIG.

was wrapped with cellophane at surgery. Over aortic area had loud "tambou
second sound (S_2), systolic ejection sound (E) and systolic murmur (SM) (upper tracing). At apex (lower tracing) first sound (S_1) not accentuated, probably due to prolongation of P-R interval. Second sound (S_2) retouched by artist to illustrate loud sound heard clinically, but photographed poorly.

at first be called a first heart sound over the aortic area is actually an ejection sound (Figs 600 601)

If the aneurysm involves the aortic valve ring, causing valvular insufficiency, a diastolic murmur may be evident in addition to the

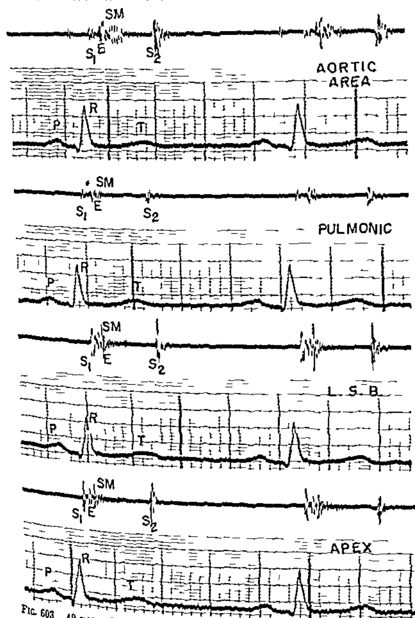
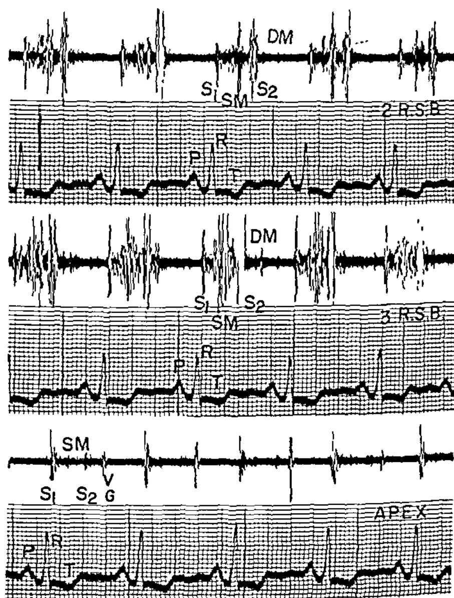


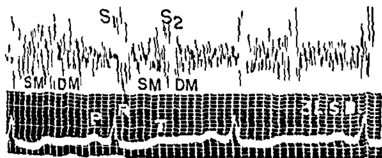
FIG. 603 49 year old man with *dissecting aneurysm of ascending aorta.* Over aortic area (upper tracing) had short systolic murmur (SM) and early systolic ejection sound (E) Systolic murmur and ejection sound heard over all areas



ventricular diastolic gallop (VG)

sion *alone, unassociated with aneurysmal* dilatation, and in others having an arteriosclerotic aneurysm of the aorta. The "tambour" type of sound is thus not specific for syphilis. In addition, an ejection sound may be noted (Figs. 600, 603), particularly over the aortic area and sometimes at the apex. Figure 600 shows the systolic murmur, aortic ejection sound and loud, accentuated aortic *second* sound. The first heart sound at the apex is diminished because of a prolonged P–R interval, which is *further evidence that* the sound which might

systolic. This is illustrated by Figures 601 and 602 In case there is rightward displacement of the aortic root, *the diastolic murmur may* be heard better along the right sternal border than the left (see discussion on Right-Sided Murmurs of Aortic Insufficiency) *In patients* with recent aortic dissection the murmurs may change At first a systolic murmur may be heard over the base of the heart, and then the appearance of a diastolic murmur suggests that the aortic valve has become involved For example we have seen a localized aneurysm in the first part of the descending aorta that resulted in a systolic murmur over the aortic and pulmonic areas Aneurysm over the abdominal aorta is *frequently* associated with a grade II to IV systolic murmur heard best over the aneurysmal site (Fig 605) In some patients *even an extensive aneurysm produces no murmur,* possibly because the sac is filled with clot and no turbulence results



A syphilitic aneurysm *of the aorta may rupture into the pulmonary* artery This is actually an arteriovenous fistula similar to patent ductus or aortic septal defect, and like them is associated with a continuous machinery murmur An example of this is shown in Figure 606 where a loud, grade V continuous murmur and a palpable thrill suddenly developed over the pulmonary area On auscultation the murmur was indistinguishable from that of patent ductus

TRAUMATIC HEART DISEASE

Trauma to the cardiovascular system may be manifest in a variety of ways The injury may be nonpenetrating from external force on the chest, such as might occur in automobile or airplane accidents or from any heavy blows against the chest wall In fact the heart may be injured from accidents and falls without a direct blow to the chest Among these injuries may be ruptures of the pericardium, myocar

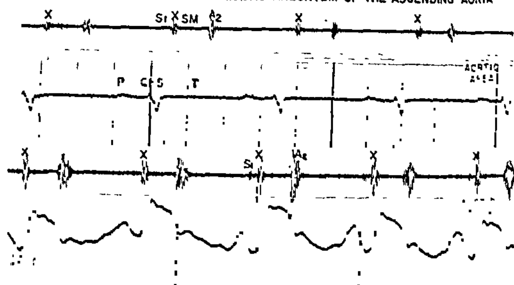


FIG. 604. 52 year old woman with syphilitic aneurysm of ascending aorta. Had faint systolic murmur (SM) over aortic area and clear early ejection sound (X). Both tracings taken over aortic area.

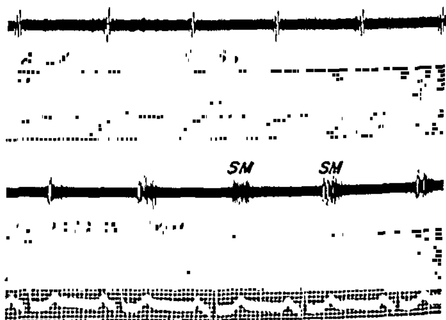


FIG. 605.
a grade IV s
mur (SM) pr
was suspected ...
suddenly.

injury other that local contusions The patient illustrated a delayed onset of symptoms which occurs in some such cases, probably from softening and necrosis and final rupture This progression may take place immediately or in hours or days In this particular instance the

' approximately six days

oted a sudden noise in his

chest which he described like a water pump leaking water around the valves He had a grade V, high pitched, musical diastolic murmur that was heard best along the left sternal border at the third interspace but was transmitted widely over the precordium Symptoms of congestive heart failure had already appeared and in view

rtic valve rupture a Hufnagel

of the descending aorta in an

insufficiency This patient was

alive three and a half years later, relatively asymptomatic but showed progressive cardiac enlargement over the years A concomitant interventricular septal defect presumably due to the trauma was also present as evidenced on cardiac catheterization prior to and after cardiac surgery

Figures 608 and 609 represent a 29 year old man who had had an automobile accident two years prior to hospitalization for a repair of an aneurysm of the first portion of the descending aorta Systolic murmurs of grade III intensity were noted over the aortic and pulmonic areas but in addition a systolic murmur was evident at the apex and along the lower sternal border The murmur along the lower left sternal border increased with inspiration suggesting the possibility of tricuspid insufficiency On cardiac catheterization tricuspid insufficiency was demonstrated and brought to mind the possibility of a traumatic rupture of the tricuspid valve as well as the aneurysm of the thoracic aorta There was no history of rheumatic or congenital

lence of penetrating injury in this case When such aneurysms are produced rupture may occur and the great majority of patients die minutes hours or days after this rupture

Occasionally patients present after trauma with symptoms or signs of pericarditis or with a clinical picture suggestive of myocardial infarction Nonpenetrating trauma can involve the pericardium setting up a pericarditis or contusion to the myocardium may injure the muscle or a coronary artery Figure 610 illustrates the auscultatory findings in a 24 year old sailor who had been injured in an air plane crash Seven months later a scratch and a clicking sound were present on auscultation which could be altered by pressure over the chest with the hand while the patient was in the sitting position These sounds were thought to be of extracardiac origin for at the time of examination a lateral x ray film still showed pleural reaction

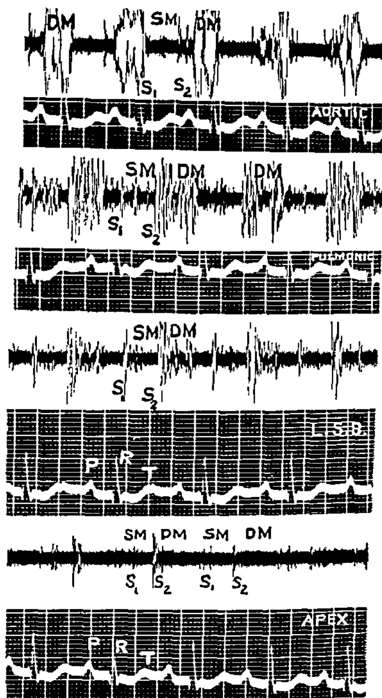
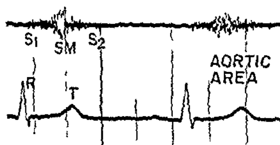


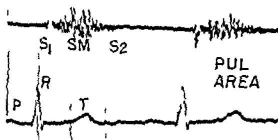
FIG. 607. 18 year old boy with traumatic rupture of aortic valve and interventricular septum (same patient as Fig. 1). Had loud grade V musical diastolic murmur (DM), loudest over aortic and pulmonic areas but also transmitted over entire precordium. In addition, had fainter systolic murmur (SM). Subsequently had Hufnagel valve inserted at surgery. Ventricular defect not repaired.

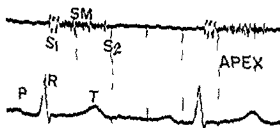
dium, interatrial or interventricular septum, valves, or various portions of the aorta.

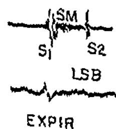
Figure 607 represents a patient who was kicked in the chest by a horse, sustaining traumatic rupture of the aortic valve and of the interventricular septum. There was no evidence of external chest

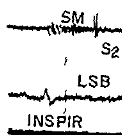
TRAUMATIC HEART DISEASE
ANEURYSM DESC THORACIC
AORTA + TRICUSPID INSUFF





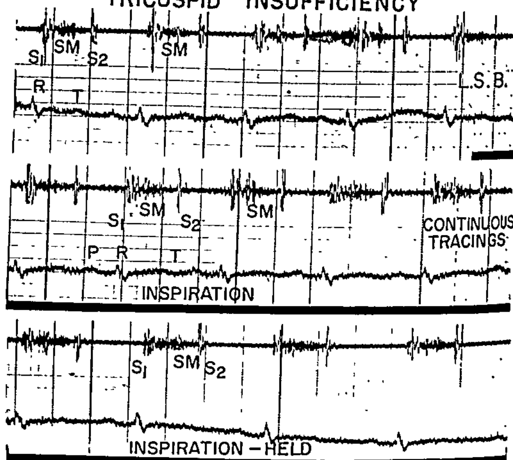






in an area adjacent to the heart. These findings subsequently dis peared, and in this case the extracardiac sounds presumably w related to trauma, particularly involving the pericardium. No e dence of valvular or septal defect was evident.

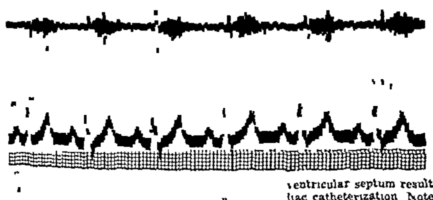
Figure 611 represents findings in an 18 year old boy who had automobile accident. The patient was unconscious and had respirato



tolic murmur (SM) along lower left sternal border which increased coincident with inspiration, as shown on continuous tracings.

distress which required a tracheotomy. Fractures of the mandible, maxilla and left humerus were present, but there was no apparent injury to the chest. Initial physical examination showed a blood pressure of 70/50, pulse 110 and no evidence of cardiac abnormality. Seven days later a precordial systolic thrill was evident, and there was a grade IV systolic murmur heard best along the left sternal border. An electrocardiogram then showed left bundle branch block, and a diagnosis of traumatic interventricular septal defect was made.

During times of war, penetrating injuries from bullets or shell fragments are common and even during peacetime such injuries occur An example of one that produced an interventricular septal defect is shown in Figure 612 Here the murmur occupied all of systole just as happens with a congenital defect.



ventricular septum result
liac catheterization Note

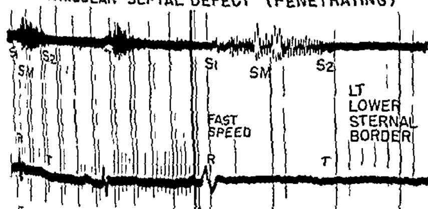


Fig. 612. Army sergeant, age 38 with injury to heart causing ventricular septal defect. Note pansystolic murmur (SM) heard at left lower sternal border Fast speed shown at right.

Traumatic Arteriovenous Fistula. Arteriovenous fistulas can occur as a result of many kinds of penetrating trauma such as stab wounds and gunshot wounds A traumatic arteriovenous fistula involving the left subclavian artery was diagnosed in a soldier wounded in Korea. A continuous machinery murmur (Fig 613) was heard in the left shoulder region and the sound simulated that of patent ductus. However the atypical location history of trauma normal

Because of deterioration in his condition cardiac surgery was performed, and an interventricular defect large enough to admit the surgeon's index finger was found. The defect was sutured and the thrill was obliterated. The patient had a stormy postoperative course,

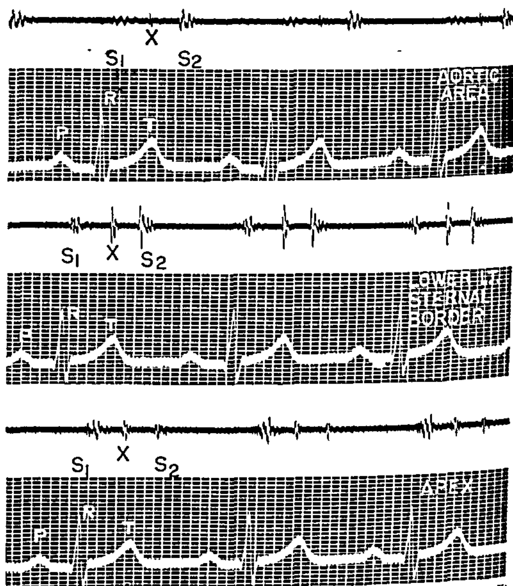


FIG. 610. 24 year old sailor injured in airplane crash. Had systolic sound (X) occurring approximately midway between first and second sounds (S_1, S_2) heard over precordium, but best over lower left sternal border (middle tracing).

and an emergency thoracotomy was necessary to correct ventricular fibrillation. The murmur and thrill reappeared, and cardiac catheterization four months later revealed evidence of ventricular septal defect. The pulmonary artery pressure was normal at that time, and the patient had no cardiovascular symptoms.

penicillin as a precaution against subacute bacterial arteritis coincident with any dental procedures infections or operations

The auscultatory features of traumatic injuries to the heart and vessels are frequently the first clues that lead the physician to suspect this complication. Promptness in diagnosis may be life saving

RUPTURE OF CHORDAE TENDINEAE

This is a relatively rare occurrence that may result from unusual exertion, trauma or infection (such as subacute bacterial endocarditis involving a chorda) It also appears to develop suddenly in some instances without any precipitating cause When rupture occurs, a pansystolic murmur of mitral insufficiency results, usually of grade IV to VI The murmur is similar to the murmur of rheumatic mitral insufficiency in that it is heard best at the apex and, if sufficiently loud, is transmitted over the precordium as well as to the left axilla and the back. The diagnosis can be made by the sudden appearance of a

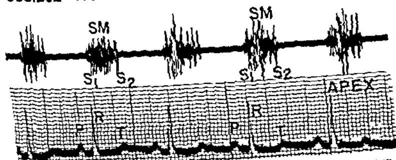


Fig. 614 A 55 year old man with a grade IV apical pansystolic murmur (SM) Patient admitted to hospital while in congestive failure Murmur had previously been noted to be grade I or II by same physician

loud murmur or a significant increase in the loudness of a previously known faint systolic murmur At times the murmur has a musical quality

One of our patients a janitor, had a known insignificant grade I or II apical systolic murmur Coincident with carrying heavy window blinds upstairs he had sudden acute and severe dyspnea A grade V pansystolic murmur with a palpable systolic thrill was now present Despite strict medical management his course was progressively downhill and he died in congestive heart failure several months later

Death may occur in a matter of hours, days or weeks after rupture of a chorda. As in the case just cited the patient may live for several months, still others live for a number of years Figure 615 illustrates the case of a 55 year old man who worked in a lumber yard where he helped to move heavy logs He had congestive heart failure when seen by his physician who for the first time noted a grade IV apical systolic murmur On previous examinations the same physician, as well

x-ray of the heart, and the absence of any previous history of a hear murmur all pointed against a diagnosis of patent ductus.

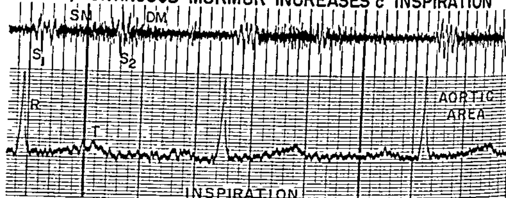
Figure 614 represents a continuous murmur heard in a 38 year ol woman. It was auscultated over the upper anterior chest and wa

TRAUMATIC ARTERIOVENOUS FISTULA



FIG. 613. Traumatic arteriovenous fistula in young soldier wounded in Korea. Had continuous machinery murmur heard over pulmonary area but loudest in left shoulder region. (Phonocardiogram taken from tape recording.)

TRAUMATIC — ARTERIOVENOUS FISTULA CHEST (BULLET WOUND) CONTINUOUS MURMUR INCREASES c̄ INSPIRATION



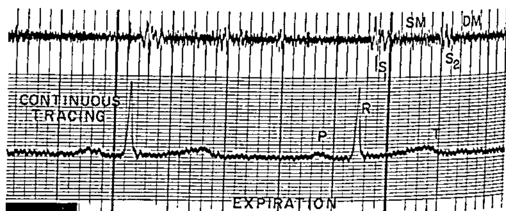


FIG. 614. Woman, age 38, with grade III continuous machinery murmur (SM-DM). Had late systolic and early diastolic accentuation of continuous murmur enveloping second soun' ... s tracing, murmur increased in ... ;ust below upper middle sternum.

loudest in the aortic area. A diagnosis of traumatic arteriovenous fistula was established when an x-ray examination of the chest revealed a bullet situated just below the upper sternum. It was then learned that the patient had been shot a number of years previously. Since she exhibited no cardiac effects or symptoms no surgery was deemed necessary. She was, however, advised take prophylactic

At the site of the myocardial infarction the normal ventricular muscle may be replaced by a thinner layer of fibrous and muscle tissue. Coincident with systole while the healthy muscle contracts, there is a paradoxical systolic bulge or expansion of this area. Symptoms and signs depend on the extent of ventricular muscle involvement in the aneurysm. Many cases are first diagnosed at postmortem examination Others are recognized on x ray or fluoroscopic examination and in still others the first clue may come from the electrocardiogram The persistence of acute changes of myocardial injury over months and years leads one to suspect the presence of a ventricular aneurysm It has been stated that there are no constant characteristic auscultatory findings of ventricular aneurysm, although a "weak" or "muffled" first heart sound gallop rhythm, systolic or diastolic murmurs or both, have been described More commonly a prominent systolic impulse is felt Congestive heart failure or peripheral embolization resulting from thrombi that form within the aneurysm may result

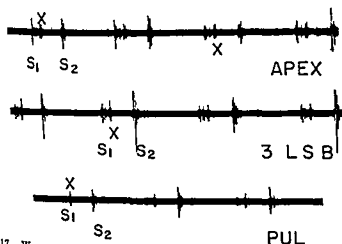


FIG 617 Woman age 50 with aneurysm of left ventricle resulting from previous myocardial infarction. Note early *systolic sound (X) heard best at apex and left sternal border* (upper and middle tracings) This sound coincided with a prominent localized systolic impulse palpated at apex

from this complication of myocardial infarction Figure 617 illustrates the case of a 50 year old woman with proven ventricular *aneurysm* as a result of previous myocardial infarction On auscultation the striking feature was the presence of an apical systolic sound in the first third of systole that was coincident with a prominent systolic impulse readily discernible by palpation over the apex This patient likewise had electrocardiographic changes as well as fluoroscopic and x ray evidence of aneurysm of the left ventricle She came to surgery for ventricular aneurysm and although this was not excised, the weakened area was reinforced by the surgeon

as others, had heard only a faint grade I or II systolic murmur. The patient responded to routine treatment of congestive failure. Two years later he was still working and his murmur was unchanged.

RUPTURE OF THE INTERVENTRICULAR SEPTUM FOLLOWING MYOCARDIAL INFARCTION

Rupture of the interventricular septum is an unusual complication following an acute myocardial infarction that involves the interventricular septum. The diagnosis, however, may be suspected in a patient with acute myocardial infarction by the sudden appearance of a loud systolic murmur, as illustrated in Figure 616. While the patient was hospitalized for treatment of acute myocardial infarction, a loud harsh systolic murmur appeared that was heard best along the lower left sternal border. The murmur was pansystolic, and, as is so often the case, death resulted a few days later from progressive cardiac decompensation. The perforation of the septum was verified at autopsy. Such a murmur is commonly associated with a thrill, and the



[illegible] septum was confirmed at postmortem [illegible] from tape recording.)

most common location is along the lower left sternal border, as in the case cited. Death usually follows in a few days to several weeks, although occasional patients have lived a few years despite the complication.

RUPTURE OF A PAPILLARY MUSCLE FOLLOWING MYOCARDIAL INFARCTION

Necrosis of heart muscle coincident with acute myocardial infarction may involve and rupture a papillary muscle, resulting in insufficiency of the mitral valve. A moderately loud systolic murmur suddenly occurs, and its features are similar to those already discussed under Rupture of Chordae Tendineae. Death in progressive congestive heart failure generally follows shortly.

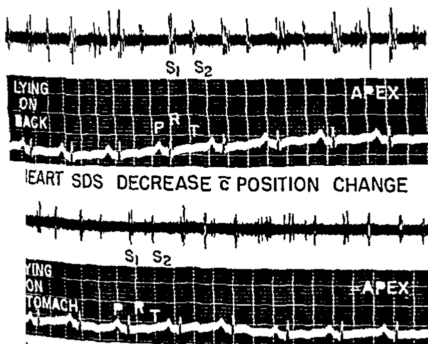
ANEURYSM OF THE LEFT VENTRICLE

This is a complication that may occur as a result of an acute myocardial infarction. The most common sites of involvement are the apex and anterior wall of the left ventricle and ventricular septum.

carcinomas. Also, Hodgkin's disease not infrequently involves the heart. The main clinical evidence of these *metastatic tumors* is the appearance of a pericardial friction or the signs and symptoms of pericardial effusion. The effusion is likely to be bloody in these circumstances.

At times a tumor involving the heart may first present with symptoms and signs of acute pericarditis with pericardial effusion, as illustrated in Figure 619. In this patient pericardial fluid was first suspected during auscultation when the patient changed his position

TUMOR OF HEART — PERICARDIAL EFFUSION



om lying on his back to lying on his abdomen and propping himself on his elbows. As the patient turned the heart sounds decreased shown on the phonocardiogram. A postmortem examination revealed the effusion to be caused by a tumor that had invaded the ricardium.

Another clue to a diagnosis of neoplastic involvement of the heart the development of paroxysmal rapid heart action or of some form f conduction defect in a patient already known to have malignant sease. A patient is recalled who had Hodgkin's disease. There was o evidence of cardiac involvement until she began to have frequent ttacks of paroxysmal atrial tachycardia. These attacks recurred as

Since this experience, patients having a history of myocardial infarction have been examined carefully for this systolic sound. Figure 618 illustrates the case of a 60 year old man with ventricular aneurysm resulting from a myocardial infarction that had occurred seven years previously. At the apex the first heart sound was slightly diminished (P–R interval was 0.20 second). A grade III, short, early systolic murmur was present, and, in addition, an inconstant systolic sound was noted in midsystole. This systolic sound occurred at the same time as a palpable impulse in a localized spot at the apex. Coincident with inspiration, as shown in Figure 618, the systolic sound decreased or disappeared; and it returned with expiration. It is felt that this sound in systole, corresponding to a localized palpable sys-

SYSTOLIC SOUND c̄ VENTRICULAR ANEURYSM
DECREASES c̄ INSPIRATION

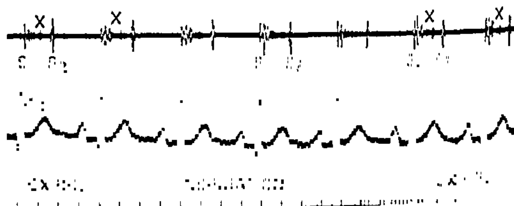


FIG. 618. Man, ... cardial infarction...

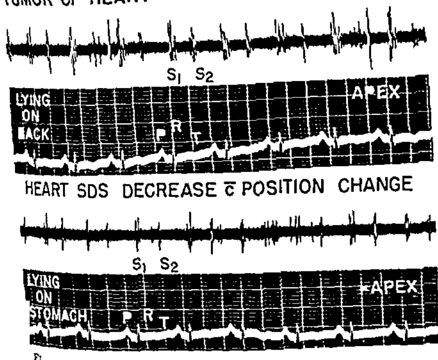
tolic impulse in a patient with a history of coronary artery disease, may be an auscultatory clue to the diagnosis of that ventricular aneurysm. The sound is most likely produced by the paradoxical systolic bulge (or expansion) of the localized aneurysm striking against the chest wall. Whether this sound is present will depend upon the location of the aneurysm, the extent of involvement and the vigor of ventricular systole.

TUMORS OF THE HEART

Tumors of the heart may be primary or secondary. The latter may occur as metastatic lesions from almost any type of malignant tumor or as extensions from neighboring structures. The most common tumors that metastasize to the heart are bronchogenic and pulmonary

carcinomas. Also, Hodgkin's disease not infrequently involves the heart. The main clinical evidence of these metastatic tumors is the appearance of a pericardial friction or the signs and symptoms of pericardial effusion The effusion is likely to be bloody in these circumstances.

At times a tumor involving the heart may first present with symptoms and signs of acute pericarditis with pericardial effusion, as illustrated in Figure 619 In this patient pericardial fluid was first suspected during auscultation when the patient changed his position



Fi

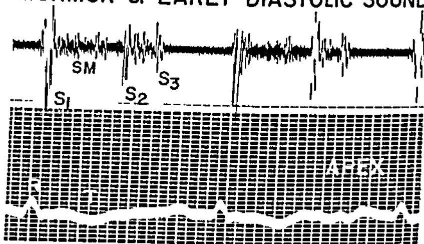
... (lower tracing)

from lying on his back to lying on his abdomen and propping himself up on his elbows As the patient turned the heart sounds decreased as shown on the phonocardiogram A postmortem examination revealed the effusion to be caused by a tumor that had invaded the pericardium

Another clue to a diagnosis of neoplastic involvement of the heart is the development of paroxysmal rapid heart action or of some form of conduction defect in a patient already known to have malignant disease. A patient is recalled who had Hodgkin s disease There was no evidence of cardiac involvement until she began to have frequent attacks of paroxysmal atrial tachycardia These attacks recurred as

frequently as several times daily and could often be arrested by carotid sinus stimulation. At postmortem examination extensive infiltration of the atrium was found. In another instance of Hodgkin's disease, the classic evidence of pericardial constriction and tamponade developed from pericardial fluid and adhesions.

A rare type of tumor producing odd auscultatory findings is illustrated by the following experience. This 42 year old nurse (Fig. 620) had had precordial pain, dyspnea on effort and paroxysmal tachycardia for three years. Evidence of pericarditis had been noted. A forceful impulse was felt in the third, fourth and fifth left intercostal spaces about 4 cm. from the sternum. A grade III apical systolic murmur was present, and a loud sound was heard in early diastole, simulating an opening snap; but do diastolic rumble was audible. The neck veins were distended and the liver was enlarged. An x-ray examination of the chest showed a 13 mm. area of radiolucency over the left lower border of the heart in the posteroanterior view. Electrocardiograms were compatible with a diagnosis of left atrial hypertrophy and epicardial ischemia. Cardiac catheterization revealed normal pulmonary and right ventricular pressure. Angiocardiograms indicated that there was a large mass posterior to the heart that was compressing the left ventricle and the left and right atria forward. An operation was performed and a tumor twice the size of the heart was found. The tumor tissue involved the pericardium and the myocardium. A line of cleavage separating the tumor from the heart was most difficult to find. The patient died during the operation, and at postmortem examination the tumor mass measured 16 x 12 x 9 cm. It



had compressed the left ventricle and projected into the left and right atrial cavities. There were metastases in the lymph nodes. The final diagnosis was neuroblastoma of the pericardium

Th... little to be done in cases of metastatic tumors of the heart. ... by ... The

main concern is to search for the rare instances in which tumors of the heart may be benign and curable by surgery Among these, myxoma of the atrium is most important.

MYXOMA OF THE ATRIUM

Benign myxoma of the atrium is rare, but with modern methods of ...agnosis and surgical treatment the lesion may be curable It occurs ...bout three times as frequently in the left atrium as in the right. ...hen present in the left atrium it generally is mistaken for mitral ...tenosis and when in the right is likely to be diagnosed as tricuspid ...tenosis or pericardial constriction No doubt in both types tumors are ...resent for a long time, possibly years, without producing signs or symptoms In some cases embolus from the tumor may be the first evidence of any abnormality

All the physical signs of mitral stenosis may be present as a result of a left atrial myxoma. There have been proved instances in which a snapping apical first heart sound, presystolic murmur and opening snap (some shown in the phonocardiogram), an accentuated pulmonary second sound, a systolic murmur and a prominent left atrium on x ray examination have all been present. In other cases fewer of

...

the operating table when the patient has been explored for mitral stenosis.

Inasmuch as a few cases of left atrial myxoma have now been diagnosed during life and an isolated one here or there has been successfully operated on, it becomes imperative to make every effort to

...

...oms (better on lying down) and changing auscultatory findings from time to time in a patient with suggestive evidence of mitral stenosis

The following case history illustrates the difficulties that may arise

This 61 year old woman (Fig 621) complained of cough and breathlessness of three years duration gradually growing worse and eventually becoming incapacitating There was no history of rheumatic fever She showed a grade II apical pansystolic murmur Many observers heard a "short sound in diastole at the apex and one observer noted a presystolic murmur The apical first sound and the pulmo-

nary second sound were accentuated. There was slight pretibial edema. The electrocardiogram showed right ventricular hypertrophy, a short P–R interval (0.12 seconds) and tall, notched P waves. Roentgen studies revealed the transverse diameter of the heart to be at the upper limits of normal and demonstrated a moderately enlarged left atrium and a slightly enlarged right atrium. The hilar vessels were moderately enlarged, and the pulmonary vasculature in general was prominent. No intracardiac calcification was seen. At operation a myxoma of the left atrium measuring 5 x 4 cm. was found that originated in the interatrial septum. Partial occlusion of the mitral orifice

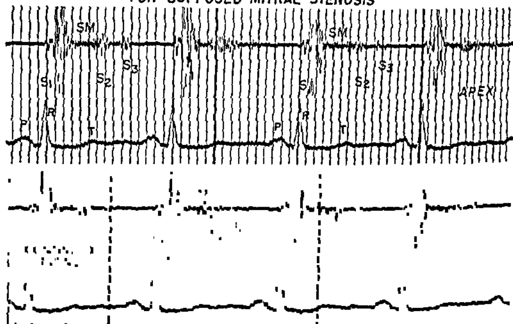


FIG. 621. A 61 year old woman with symptoms of cough and breathlessness of three years' duration; had progressive incapacitation. No history of rheumatic fever. Mitral stenosis suspected because of accentuated first sound (S_1) at apex and a third sound in diastole (S_3) simulating opening snap; also had grade II systolic murmur (SM). At operation, myxoma of left atrium found; no mitral stenosis. Note variation of systolic murmur (SM) from beat to beat. Had short P–R interval (0.11 second), accounting for loud first sound

was present. The accentuated first sound could be explained by the short P–R interval. The systolic murmur was noted to vary considerably from beat to beat. Similarly, the third sound showed minor variations in timing after the second sound and also variations in intensity. We have noted similar variations in the third sound in the phonocardiograms of another published report of a case of atrial myxoma. It might prove helpful to look for these variations in the systolic murmur and in the timing of the third heart sound when considering the diagnosis of myxoma of the atrium.

The diagnosis should be suspected if a patient with symptoms resembling mitral stenosis has spells of dyspnea, palpitation or faint-

ness that are relieved by lying down In most cardiac patients these symptoms especially breathlessness, are worse in the recumbent position Another possible clue to the diagnosis is the sudden development of an arterial embolus either in a patient who has no good evidence of heart disease at all or in a so-called mitral case with a normal sinus rhythm Whenever the possibility of an atrial myxoma comes to mind one should seriously consider doing angiocardiograms or possibly cardiac exploration. The former procedure is more easily done for lesions of the right atrium than the left If a surgeon encounters a myxoma while operating for mitral stenosis it is best to retreat and plan to operate at a later time either under hypothermia or with open heart surgery

AUSCULTATORY FINDINGS IN HYPERTENSION AND HYPERTENSIVE HEART DISEASE

In patients with hypertension a systolic murmur is frequently heard over the aortic or apical areas, varying in intensity between grade I and III (Figs 622, 623) The apical systolic murmur is most likely due to a relative mitral insufficiency coincident with a large left ventricle. The murmur over the aortic area probably results from a dynamic dilatation of the first portion of the ascending aorta just beyond the aortic valve This represents a relative type of aortic stenosis and at postmortem examination no abnormality whatsoever may be noted in this region As already discussed under Unusual Etiologies of Aortic Insufficiency patients with hypertension may also have diastolic murmurs of aortic insufficiency Such a murmur is likened to the murmur of insufficiency of the pulmonary valve coincident with significant elevation of pulmonary artery pressure When arterial hypertension results in aortic insufficiency reduction of diastolic blood pressure to readings around 90 to 100 may cause this murmur to disappear It may reappear with the return of high diastolic readings.

The Auscultatory Sounds of Hypertension The aortic component of the second heart sound is accentuated with hypertension (Figs 622 623) and the intensity generally corresponds to the degree of diastolic pressure elevation In addition in some cases of hypertension over the aortic area an ejection sound is heard in early systole Such an ejection sound may occasionally be heard at the apex A frequent finding in patients with hypertension is an atrial sound or atrial gallop in presystole (Fig 622) Such a sound does not necessarily connote the presence of congestive heart failure as it would if the ventricular type of diastolic gallop were present. The atrial sound is heard best along the lower left sternal border and at the apex, and in those cases having associated congestive heart failure a ventricular type of diastolic gallop may be evident. The atrial gallop is often mistaken for a split first heart sound

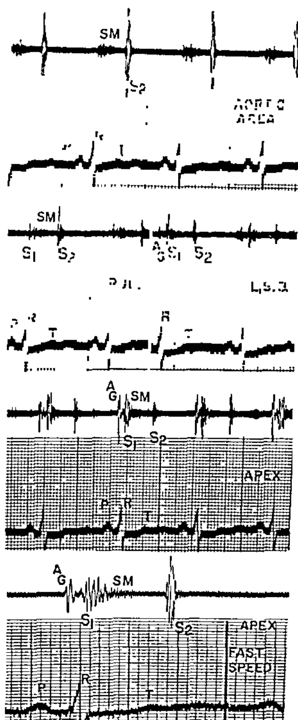


FIG. 622. Woman, age 68, with hypertension and hypertensive heart disease (blood pressure 260/140). Over aortic area (upper strip) note systolic murmur (SM) and accentuated second sound (S_2). A systolic murmur (SM) was also heard along sternal border and at apex. Note prominent atrial gallop (AG) heard best at apex (lower two tracings).

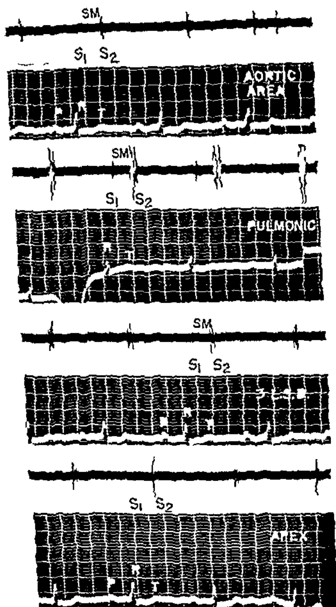


FIG. 623. Patient with *malignant hypertension (blood pressure 230/150)* who died six weeks after this recording was taken. Had faint systolic murmur (SM) over aortic and pulmonic areas and *along left sternal* border. Second sound (S_2) accentuated, and split was heard best over pulmonic area (second strip)

CHRONIC COR PULMONALE

This form of heart disease results from pulmonary disease, usually of long standing. The most frequent cause is chronic pulmonary emphysema. Patients with chronic cor pulmonale often have an emphysematous chest with an increased anteroposterior diameter. Abdominal breathing is evident. Cyanosis may also be present, and for this

SEVERE EMPHYSEMA—CHRONIC COR PULMONALE—HEART SDS. VERY FAINT OVER PRECORDIUM-WELL HEARD OVER XIPHOID & EPIGASTRIUM

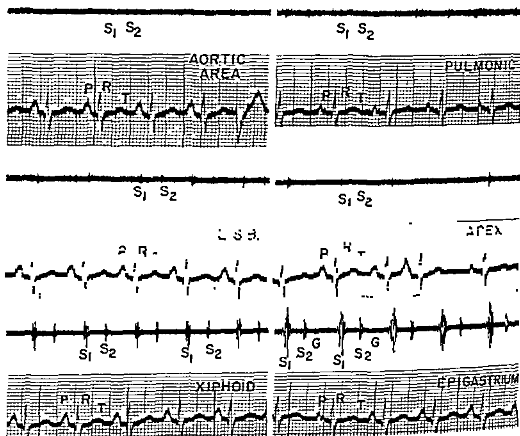


FIG. 624. 69 year old man with severe emphysema and chronic cor pulmonale. Note faint heart sounds (S_1, S_2), over usual cardiac areas (upper and middle sections). Sounds well heard over xiphoid area (left lower strip) and epigastrium (right lower). Also note ventricular diastolic gallop (G) heard over epigastrium but not at apex.

reason confusion with congenital heart disease sometimes results. Clubbing of fingers and toes is uncommon, although it does occur. On auscultation, both heart sounds over the precordium are apt to be faint. This is due to the interposed emphysematous lung tissue, for actually the sounds are not decreased. Often they can be heard well if one listens over the xiphoid area or just below the costal margin in the upper middle or left epigastrium (Fig. 624). Over the precordium, the sounds are generally heard best along the lower left

sternal border, particularly with the patient in an upright sitting position and leaning forward The sounds are not always diminished, especially if the anteroposterior diameter of the chest is not greatly increased The second sound is variable At times it is greatly accentuated and closely split. Gallop rhythm is not uncommon, and a ventricular diastolic gallop is heard in the presence of failure

Figure 625 illustrates a 28 year old man with chronic cor pulmonale A diagnosis of congenital heart disease had been suspected

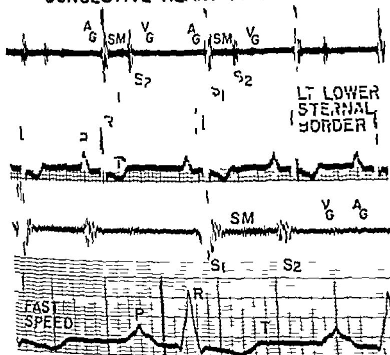


FIG. 625 ... or pulmonale. Had been suspected of cyanosis Heart sounds (S_1 S_2) heard ...dition both atrial (AG) and ventricular ...racing shows phonocardiogram at fast speed at same area

because some cyanosis was present. Right ventricular hypertrophy and a large P pulmonale were evident on the electrocardiogram The x ray film showed a large pulmonary artery right ventricular hypertrophy and vascular markings that appeared to be slightly increased A moderate pectus excavatum of the cup type was present and the jugular venous pulse was accentuated A grade II apical systolic murmur was present, the first and second sounds at the apex were diminished a gallop rhythm was heard resulting from both atrial

and ventricular diastolic gallops. Over the pulmonary area, however, the second sound was accentuated and closely split. The splitting increased slightly on inspiration. This patient had a long-standing history of chronic pulmonary disease, and now had chronic cor pulmonale with congestive heart failure. Symptoms of cardiac decompensation were evident, and he responded well to measures for treatment of congestive heart failure. In most of these cases, however, the ultimate prognosis is poor.

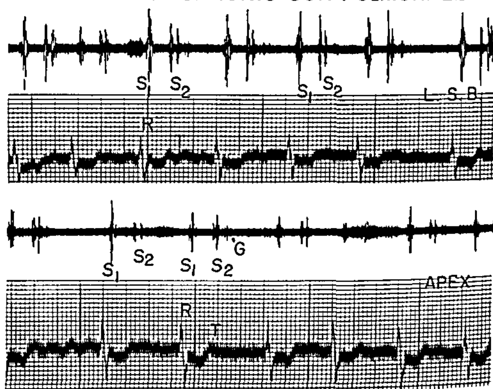
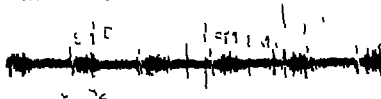


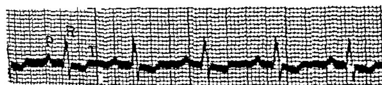
FIG. 626. 45 year old man During life, differentiation could not be made between
(1) primary pulmonary
cor pulmonale resulting
the latter. At left stern
sound (S_2) widely split.
ing right ventricular ou
that seen in pulmonic stenosis At apex (lower strip) note ventricular diastolic gallop (G).

Recurrent pulmonary emboli may produce severe pulmonary hypertension with resultant chronic cor pulmonale. This condition is frequently unrecognized, and the patient is regarded as suffering from primary pulmonary hypertension. It may be impossible to distinguish the one condition from the other, and because of this it is wise to treat such patients with anticoagulants. The patient shown in Figure 626 illustrates this problem (see discussion under Primary Pulmonary Hypertension).

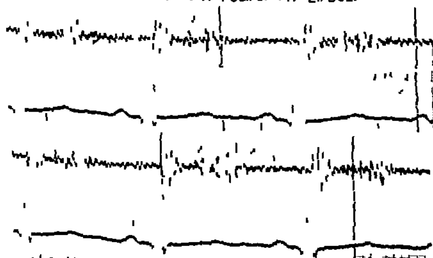
PULMONARY ARTERIOVENOUS COMMUNICATION FOLLOWING RECURRENT PULMONARY EMBOLI

Figures 627 and 628 represent two patients both with proven chronic venous thrombosis, who had recurrent pulmonary emboli

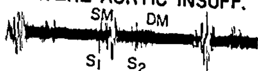




months later



... 53 year old man with thrombophlebitis (lower extremities) and recurrent pulmonary emboli. Previous hospitalizations on several occasions had not revealed continuous murmur. On last admission a grade III continuous murmur (SM-DM) heard over pulmonic area. Note late systolic accentuation (SM) and diastolic component (DM).



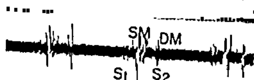
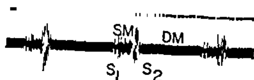


FIG. 629. 52 year old man with rhe matoid spondylitis. Had all classic fe tures of severe aortic insufficiency. Sy

der.





Chronic cor pulmonale with cardiac decompensation resulted in both patients. Each had had previous hospitalizations and numerous examinations in the same hospital, often by the same physician. A continuous murmur with systolic accentuation developed over an area of the lung in each patient. Figure 627 shows the murmur over the right lung base, and in Figure 628 the continuous murmur was loudest over the pulmonary area. Both patients had pulmonary hypertension as a

result of the recurrent emboli to their lungs Possibly in such cases the continuous murmur was related to a pulmonary arteriovenous communication that became evident as pulmonary hyperten on developed or the murmur was produced at the site of a partially occluded arterial vessel

ARTHRITIS AND HEART DISEASE

The association between arthritis and heart disease is not clear In some instances there appears to be a direct relationship rheu

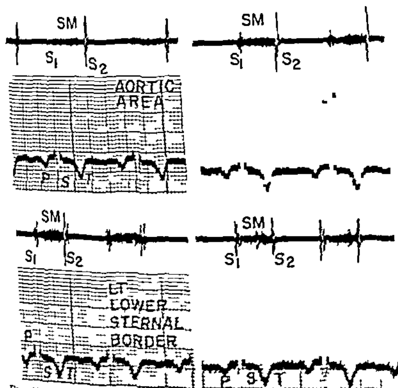


FIG. 630 A 52 year old man with rheumatoid spondylitis and mitral insufficiency Note grade III to IV pansystolic murmur (SM) heard best along lower left sternal border and at apex.

matoid spondylitis and aortic insufficiency is an example of this (Fig 629) Although aortic insufficiency is more common other valvular involvement may be present, as illustrated by the case of the patient in Figure 630 an instance of rheumatoid spondylitis and mitral insufficiency

Rheumatoid arthritis is sometimes associated with valvular lesions either mitral or aortic. Figure 631 illustrates a combination of rheu-

matoid arthritis and mitral insufficiency. A patient currently being examined regularly in our cardiac clinic who had a previous mitral commissurotomy has disabling rheumatoid arthritis as her present chief complaint.

None of the three patients cited here as examples of arthritis associated with heart disease had a history of rheumatic fever.

MYOCARDITIS

This category of heart disease is comprised of a large number of conditions which primarily affect the myocardium. These include virus or bacterial infections, so-called collagen diseases such as lupus and periarteritis, beriberi, thyrotoxicosis, amyloidosis, hemochromatosis, toxic myocarditis from various drugs, fibroelastosis, glycogen storage disease, sarcoidosis and Fiedler's myocarditis. The result may

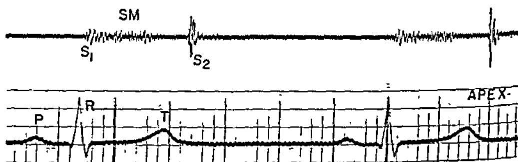


FIG. 631. Woman, age 35, with rheumatoid arthritis and grade III apical systolic murmur (SM). Known murmur present for 14 years. No history of rheumatic fever.

be angina, congestive heart failure, conduction defects, arrhythmias and cardiac enlargement. Gallop rhythm is a frequent finding in patients with myocarditis, particularly when congestive heart failure is present (Figs. 632 through 639). Often both atrial and ventricular gallops are present, or there may be a summation gallop. At other times, the auscultatory findings of right or left bundle branch block may be noted. The pulmonic component of the second heart sound may be accentuated if failure is present. A systolic murmur (grade II or louder) is commonly found at the apex, along the sternal border, or over the pulmonic or aortic regions.

The diagnosis of myocarditis should be suspected in a patient who has a ventricular diastolic gallop, cardiac enlargement and other symptoms or signs of heart failure that cannot be explained by the more usual causes. This is illustrated by Figures 632 through 639. Two patients with proven lupus erythematosis had prominent ventricular diastolic gallops, as shown in Figures 632 aud 633. One patient (Fig. 632) also had a delayed P–R interval and, at times, a sum-

LUPUS MYOCARDITIS — AURICULAR AND VENTRICULAR DIAST GALLOP

SM DM

V_G A_G S_2 S_1 S_2 V_G A_G

APEX

R T P R T P

SM DM

S_1 S_2 V_G A_G S_1

APEX FAST SPEED

T P R T P

FIG. 63? 3 year old man with proven lupus erythematosis Had chronic congestive heart failure and died almost three years later At apex had both systolic (SM) and diastolic (DM) murmurs and in addition had both ventricular (VG) and atrial (AG) diastolic gallops. Lower tracing shows phonocardiogram at faster speed

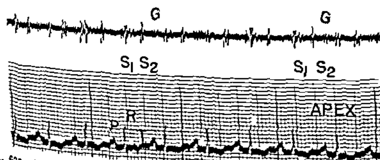
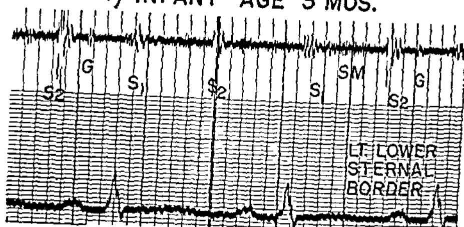


FIG. 633. Patient with proven lupus erythematosis. Note diastolic gallop (G) at apex heard in addition to first and second sounds (S_1 S_2)

mation gallop that was produced by the superimposed ventricular and atrial gallops. A diastolic rumble was present at other times. This patient had been receiving steroid therapy for several years, and although he suffered from chronic failure his disease was fairly well controlled by this medication and by measures to control heart failure.



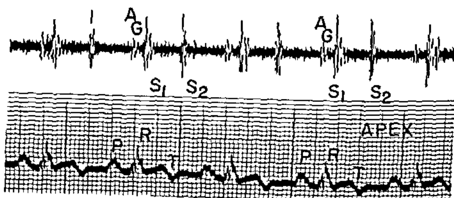


FIG. 634 Two patients with clinical diagnosis [illegible] ... [illegible] revealed no evidence of shunt or specific congenital anomaly. Note atrial gallop (AG) well heard at apex

He died approximately two years after the date of the recording shown in Figure 632.

Another patient (Fig. 634, lower tracing), a 25 year old man, was evaluated for possible congenital heart disease because of the finding of cardiomegaly, prominent atrial gallop and a bundle branch block. A systolic murmur (grade II to III) was heard at the apex, and the patient had a history of some kind of "heart condition" since birth. A cardiac catheterization revealed no evidence of a shunt or of any

other congenital anomaly It was thought that the most likely diagnosis was fibroelastosis

Most patients with fibroelastosis die during the first years of life Figure 634 (upper tracing) illustrates the case of a three month old baby girl who had congestive heart failure, gallop rhythm, rales and an enlarged live ... ponded well to treatment with digitalis, penicillin and ox ...
grade fever of o ...
failure followed The phonocardiogram shown in Figure 634 was taken two weeks after this acute episode A grade II systolic murmur

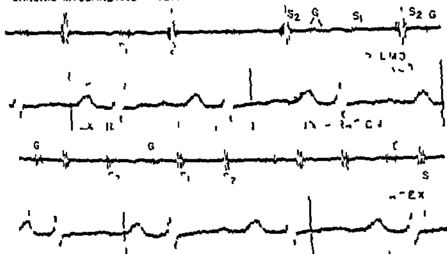
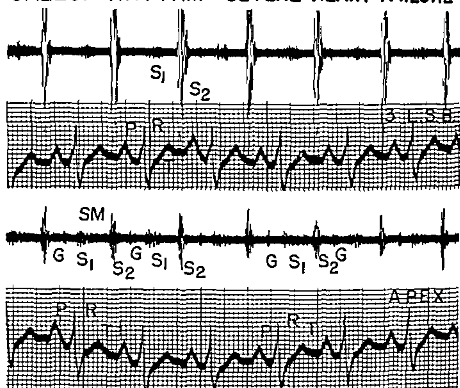


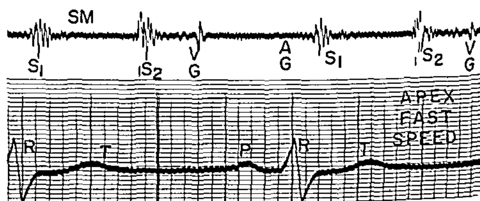
Fig 635
ion. Previou
l diastolic
controlled on
ops plus di

and a diastolic gallop rhythm were the only significant auscultatory findings Cardiomegaly was already present and the prognosis was considered poor

Figures 635 and 636 illustrates examples of idiopathic myocarditis Both of these patients had chronic cardiac decompensation atrial and ventricular diastolic gallops and at times a diastolic rumble in addition to a systolic murmur Both patients had previously been diagnosed as having rheumatic heart disease, and in one patient (Fig 635) an operation for mitral stenosis had been contemplated

Two patients with hyperthyroidism and clinical evidence of cardiac decompensation are shown in Figure 637 Examples of myocarditis





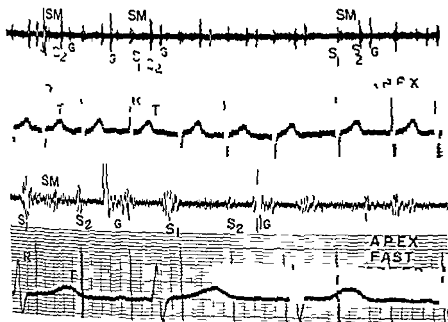
tracing). Lower tracing showing both atrial (AG) and ventricular (VG) gallops heard on prior hospitalization 15 months previous. Death occurred three months after second hospitalization. Postmortem revealed chronic myocarditis; etiology undetermined.

associated with sarcoidosis, Wegner's granulomatosis and eosinophilic leukemia are shown in Figures 638 and 639.

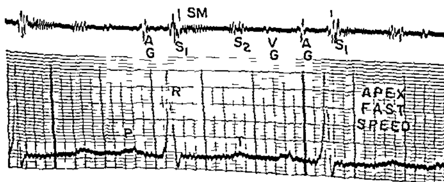
Murmurs, particularly systolic, may be a prominent auscultatory finding leading to a mistaken diagnosis of a primary valvular or congenital defect, such as mitral insufficiency, ventricular septal defect

and occasionally aortic stenosis Sometimes a diastolic rumble is heard, leading to confusion with mitral stenosis A recent patient had a grade III to IV pansystolic murmur (heard best at the lower sternal border and transmitted to the left axilla, the lung bases and the lower interscapular area), but no evidence of a shunt was found on right and left heart catheterization, and there was no other evi-

HYPERTHYROIDISM — AUR FIBRILLATION AND VENT DIAST GALLOP



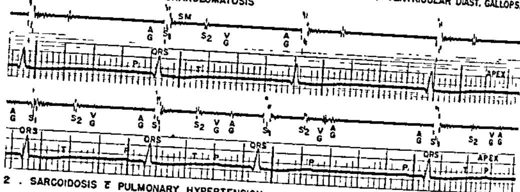
ANOTHER PT — HYPERTHYROIDISM — REGULAR RHYTHM AUR & VENT DIAST GALLOPS



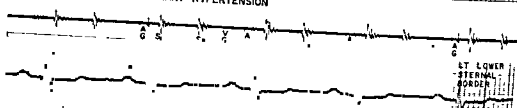
(AG) diastolic gallops

2 PATIENTS ē MYOCARDITIS OF UNUSUAL ETIOLOGY – AURICULAR & VENTRICULAR DIAST. GALLOPS.

1. WEGNER'S NECROTIZING GRANULOMATOSIS



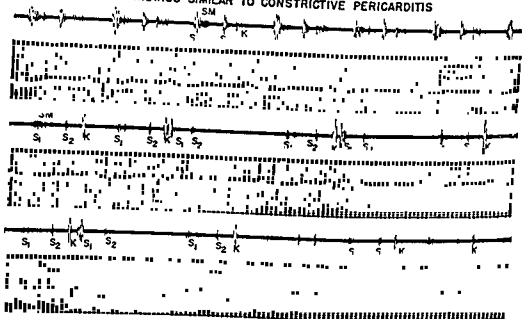
2. SARCOIDOSIS ē PULMONARY HYPERTENSION



and ventr ... usual etiology, both having atrial
Wegner's ... acings: 44 year old woman with
tricular (VG) ...

... patient with sarcoidosis and pulmonary hypertension. Note both atrial (AG) and ventricular (VG) diastolic gallops in addition to heart sounds (S_1, S_2).

EOSINOPHILIC LEUKEMIA INVOLVING HEART — LOUD DIASTOLIC SOUND AT APEX — CARDIAC CATH. FINDINGS SIMILAR TO CONSTRICTIVE PERICARDITIS



... sound (K) heard along left sternal border and apex. Systolic murmur (SM) also present at lower left sternal border and apex (second and third tracings). Ventricular extrasystoles denoted by X.

dence of a primary valvular lesion A ventricular diastolic gallop was also present at the apex. The presumable diagnosis was fibroelastosis involving the myocardium and possibly the mitral and/or tricuspid valves causing the systolic murmur of valvular insufficiency

PULSUS ALTERNANS AND ALTERNATION OF HEART SOUNDS OR MURMURS

Pulsus alternans is a mechanism to be noted in the peripheral pulse in which the strength or volume of the pulse alternates from cycle to cycle (Fig 640) This occurs in a regularly beating heart and is independent of respiration It is most commonly found in hypertensive and coronary artery disease, but also in valvular disease, especially aortic stenosis It is thought to be due to impairment in contractility of the heart muscle It can be detected on palpation of the radial pulse and is readily portrayed if a pulse tracing is taken The most common and convenient method for its detection is while determining the blood pressure If the pressure is maintained at the



Fig 640 Schema of pulsus alternans in tracing of radial pulse. Note that pulse is regular but larger waves are present with each alternate beat.

moment the first sounds are heard below the sphygmomanometer cuff they will be found to alternate in intensity When the condition is very marked only the stronger beat will be heard at the top systolic level, and at a point 10 mm. or more lower both beats will be audible displaying alternation in intensity (Fig 641) It must be appreciated that with the usual pulsus alternans both *sounds* are heard at the upper systolic level but alternate in intensity Having the patient momentarily stop breathing (but not strain) will aid in the detection of the alternation Pulsus alternans may be detected by palpation of the radial or femoral artery In its early development, alternation may be present only for several seconds after a premature beat As is well known this disturbance has a rather grave prognostic significance It indicates serious myocardial disease although there are some patients who live a considerable number of years after it is first detected When it is found during a paroxysmal tachycardia or whenever the rate is very rapid and regular (Fig 642) the prognosis need not be grave at all

What has not been sufficiently appreciated is that in many cases alternation of the strength of ventricular contraction can be heard over the precordium (Fig 643) This will be evident in alternation

of the intensity of heart sounds or of any accompanying murmur (Fig. 644). In one of these cases, striking alternation was present in the second sound because the patient had marked hypertension and the first sound was very faint. Such alternation of murmurs has

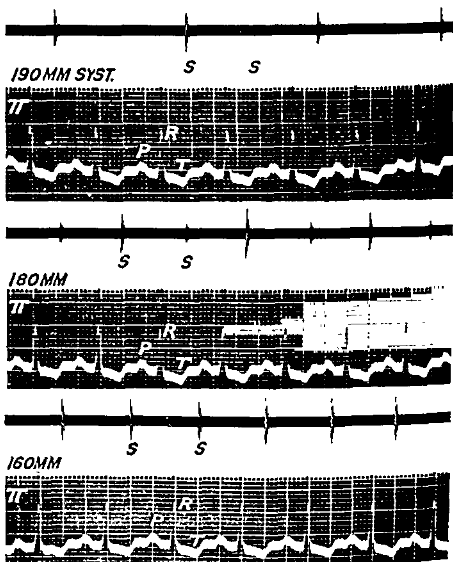


FIG. 641. A 60 year old man with hypertensive heart disease and congestive failure. Pulsis alternans (S) was noted with the blood pressure cuff. Note marked alternation (S) present at 190 mm. systolic (upper tracing). In fact, each alternate sound was practically inaudible. At 180 mm. (middle tracing) the alternation was still quite marked, while at 160 mm. the sounds [illegible] intensity. Alternation of the second sound was [illegible] (see Fig. 643). The sound tracings were obtained [illegible]

appeared to be more distinct on careful auscultation than in the phonocardiograms. When this finding is detected, it has the same general grave prognostic significance that peripheral pulsus alternans entails. In fact alternation of the ventricle can be detected at times

by inspecting or palpating the apex impulse. We recently saw such a case with conspicuous visible alternation of the apex impulse and the carotid pulsation Also, alternation of strength of ventricular contraction may be seen under fluoroscopic examination of the heart.

Certain precautions are necessary in the interpretation of alternation With inspiration both the peripheral pulse and heart sounds

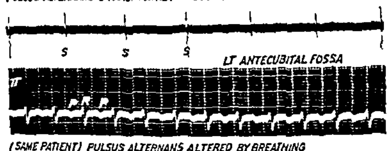


FIG. 642. Woman 54 years of age with thyrotoxicosis. Blood pressure tracings. Note regular alternation of sounds (S) heard at left antecubital fossa (upper tracing) At other times the alternation was not as marked and showed a superimposed pulsus parodoxus (lower tracing) In this instance alternation was present and also sounds (S) became quite faint or absent with inspiration.

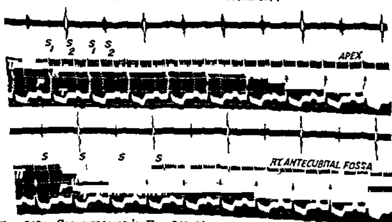
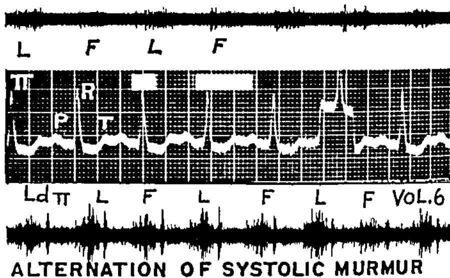


FIG 643. Same case as in Fig 641 Note alternation of second heart sound (S_2) heard at apex Also note faint S_1 with slightly prolonged P–R interval (0 22 second) Pulsus alternans (S) also present with blood pressure cuff (lower tracing)

may decrease slightly. This may not only produce a pseudoalternation (Fig. 645), but may disturb the true alternation so that it may be overlooked (Fig. 642, lower tracing). This is particularly true



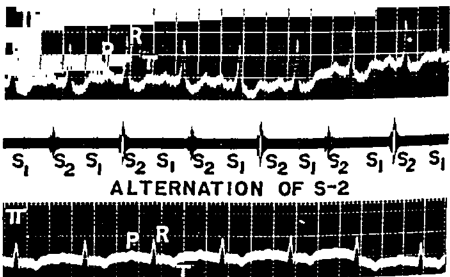
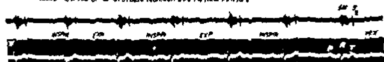


FIG. 644. Upper tracing: Woman, age 57, with aortic stenosis. Alternation of systolic murmur, loud (L) and faint (F), better heard than photographed. Middle tracing: A 68 year old woman with calcific aortic stenosis. Definite pulsus alternans present at brachial artery and alternation of aortic systolic murmur, loud (L) and faint (F), present. Lowest tracing: Same case as Fig. 641.

when the pulse pressure is small, for with each inspiration the peripheral pulse is sufficiently decreased to diminish the volume of even the larger of the two alternating beats. If the respiratory rate happens to be about one-half the heart rate, every other peripheral pulse may be smaller and every other heart sound appear to be fainter.

This is wel
46 and the ...
alternated in intensity, but this was due to the ...
In fact, one could have suspected that alternation was not present,
as it is almost never detected with a slow heart rate Another precau-
...cles must be identical—when the
...rate beats vary slightly in dura-
tion ... (Fig 646)

A striking illustration of pulsus alternans is shown in Figure 641
The alternation was so marked at a blood pressure level of 190 mm
that ... and at 180 mm the weaker
beat w...
clear ...



tion (exp). The heart rate was 46 and respiratory ...



This represents therefore a pseudoalternation for in true alternation the cycles must be of equal strength

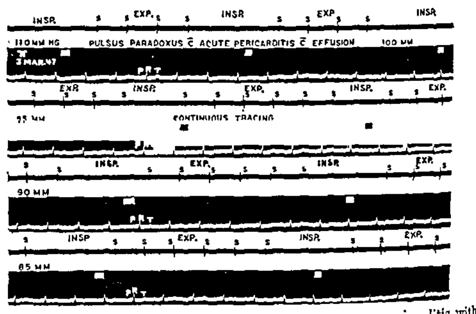
As a rule, the presence of definite alternation of the pulse connotes
... and with it a ventricular diastolic gallop is
...
are and appear before the onset of rales and edema

PULSUS PARADOXUS

Pulsus paradoxus consists of the disappearance or the *marked* decrease of the volume of the peripheral pulse with inspiration It is regarded as a sign of pericardial effusion or of constrictive pericarditis However it can frequently be observed in extremely ill patients with a thin pulse and a low blood pressure who are suffering from a variety of conditions cardiac and noncardiac in nature It is

common in asthmatic states and emphysema. This sign can readily be detected on palpation of the radial pulse and also by observation of the loudness of the sounds while determining the blood pressure. In the case illustrated by Figure 647, the pulse decreased strikingly with each inspiration. This is well shown at systolic blood pressure levels of 110 to 85 mm. This patient had a marked pericardial effusion, and when the effusion had disappeared the pulsus paradoxus was, for the most part, absent (Fig. 648).

Other factors besides the heart must play a role in the production of pulsus paradoxus. Apart from the momentary decrease in cardiac output that may occur with each deep inspiration, the movement of

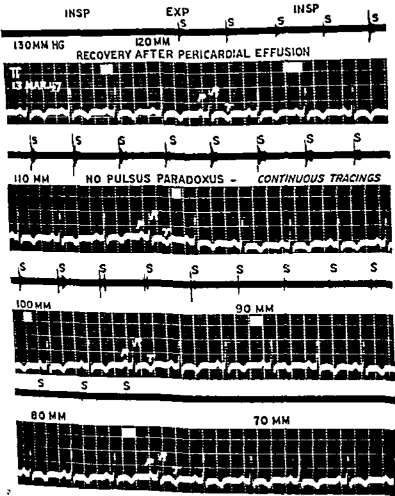


the systolic level.

the clavicle compressing the subclavian artery may have a profound effect upon the pulse. This is well shown in Figure 649. Here, in a normal individual, the radial pulse disappeared as a result of a voluntary backward motion of the shoulder, though respiration continued. No doubt patients who are gravely ill with heart failure and who are struggling for air may be partly compressing the subclavian arteries with each inspiration. Even normal individuals may show slight or marked pulsus paradoxus without deliberately throwing the clavicle backward (Fig. 650). Although it is evident that pulsus paradoxus is present in a variety of conditions, when it is marked and detectable over a wide range of pressure it is suggestive of pericardial effusion or constriction.

PISTOL SHOT AND DUROZIEZ'S SIGN

In cases of aortic insufficiency, there is often heard in peripheral arteries especially the femoral arteries, an explosive sound called the pistol shot (Fig 651) A similar sound though generally less pronounced, may also be present in other conditions such as hyper-



... phenomenon.

...yroidism and anemia, in which the heart beat is hyperactive and ...e pulse pressure increased When pressure is exerted with a stetho...ope over a large vessel like the femoral artery, a systolic murmur ...adily becomes audible In cases of aortic insufficiency however ...hen firm pressure is applied, a diastolic murmur may be heard as ...ell (Fig 651) This is called Duroziez s sign The same violent con

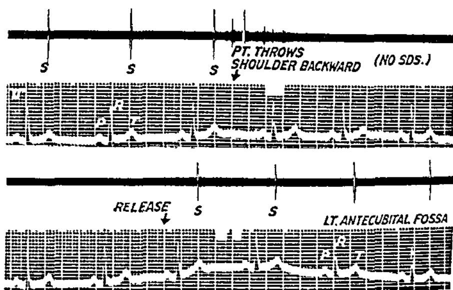


FIG. 649. Man, age 57, with no heart disease. Continuous blood pressure tracing,

cedure, and the radial pulse disappeared at the same time as the sounds.

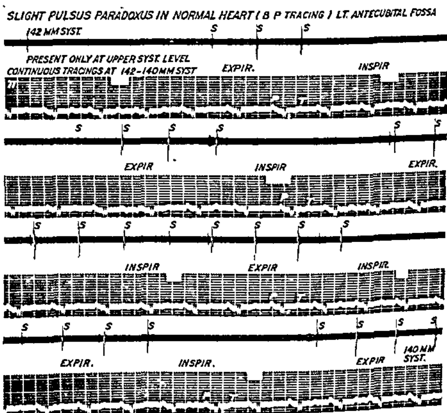


FIG. 650. A 44 year old man with essentially normal heart. Continuous blood pressure tracing (142–140 mm. systolic), left antecubital fosa Pulsus paradoxus present at upper systolic levels over a very narrow range. Sounds (S) absent with inspiration, present with expiration, not present at lower levels. This phenomenon may be present in normal individuals.

traction that produces the pistol shot (ejection sound) and other characteristics of aortic insufficiency, such as the Corrigan pulse or capillary pulse also can cause a loud snapping sound in early systole that can be heard below the clavicles (Fig 652) At times this may be confused with the heart sounds, and may even lead the observer to believe that a gallop rhythm is present (Fig 302)

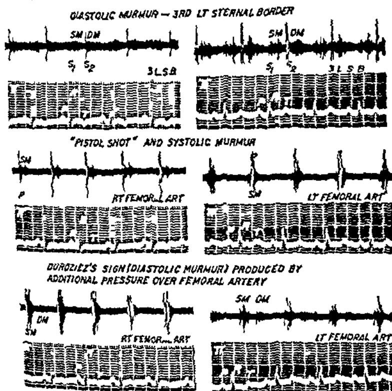


Fig 651 Two cases of marked aortic insufficiency Tracings on the left Man

e-e clearly audible This is Duroziez's sign.

BLOOD PRESSURE OBSERVATIONS

There are some simple aspects of determination of blood pressure that are often overlooked by the general practitioner One ordinarily inflates the cuff to above the point where sounds are no longer audible then determines the point at which sounds are first heard (systolic) and then when they either disappear entirely or markedly change in intensity (diastolic) (Fig 653) It should become a routine habit

when determining the blood pressure, to try to detect pulsus alternans as previously described. In making the first determination in a new patient, it is well to feel the pulse at the same time that the systolic reading is being obtained. This will avoid several errors that might otherwise arise. We saw an instance in which a family physican stated he could not obtain a blood pressure reading in a young girl who complained of headaches. It was found that she had extreme hypertension with a reading of about 270/175. The physician had always inflated the cuff to about 150 to 160 and never heard any sounds, since they were below the diastolic level. Had he felt the

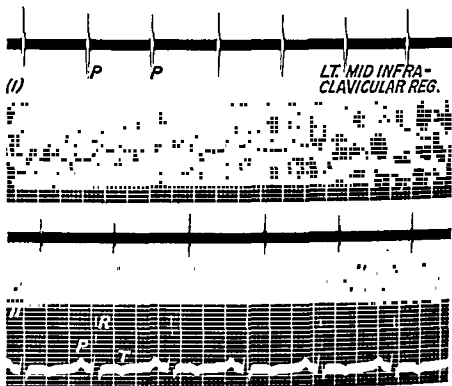


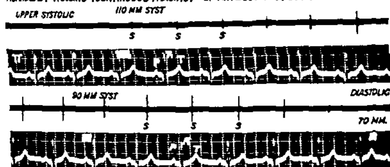
FIG. 652. Two cases with marked aortic insufficiency. In each case a loud "pistol-shot" or "ejection" sound (P) was heard bilaterally in the region of the infraclavicular fossae. Note that (P) in each case occurs in approximate midsystole.

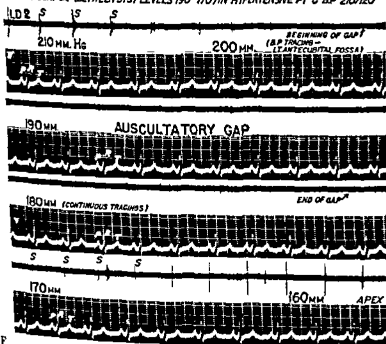
radial pulse he would have known that the systolic level was a good deal higher, and he would have avoided the error.

Another pitfall is due to beginning the determination at the so-called auscultatory gap. There are times when the sounds come through at a certain level, only to disappear for 10 or more mm. as the pressure is lowered, then returning before the diastolic level is reached. In the case illustrated in Figure 654 a reading of 170/120 was obtained by one physician when the true figure was 210/120. This mistake would not have been made if the observer had felt the radial pulse. At a level of 190 to 180 when he heard no sounds below the blood pressure cuff, if he had felt the pulse, he would have found it

present and would quickly have increased the pressure and discovered his error

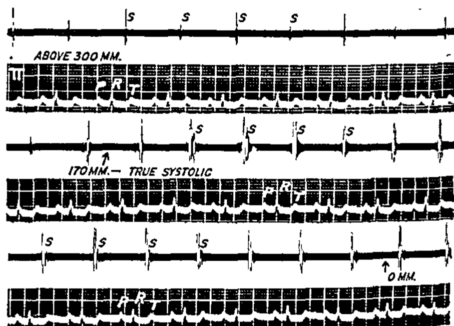
There is another uncommon error that may arise in the determination of the blood pressure. When the peripheral pulse is very explosive, as in some cases of aortic insufficiency, a violent sound will





F ... at 170 mm. (lowest tracing) This patient had been seen one-half hour previously in another clinic where the blood pressure had been erroneously recorded as 170/120

be made as the blood stream strikes the top of the inflated cuff. This sound may be transmitted to the arm just below the cuff and be audible no matter what degree of pressure is employed. We once saw a young girl of 15 whose physician was alarmed because her blood pressure was extremely high. On examination she was found to have free aortic insufficiency and a marked Corrigan pulse. On ordinary estimation of the blood pressure, sounds were clearly audible even beyond the level of 300 mm. The pulse, however, was first felt at 150 mm. This was the true systolic reading, though the sounds were pres-



ent all the way down to zero. Here, also, the palpatatory method would have avoided the difficulty. A similar experience is illustrated in Figure 654. The presence of sounds in aortic insufficiency when the cuff is not inflated at all is shown in Figures 655 and 656.

There are frequent instances when, for one reason or another, it is difficult or impossible to hear the sounds below the sphygmomanometric cuff. Under these circumstances a systolic reading may still be obtainable by palpating the radial pulse.

Other difficulties arise when the heart rhythm is irregular. In the presence of atrial fibrillation, there is no simple satisfactory method of determining the pressure levels. Inasmuch as the beats vary in strength, it becomes a complicated problem in dynamics to measure

CONT NUOUS B.P TRACING OF PATIENT C̄ RHEUMATIC AORTIC INSUFFICIENCY
B.P 162/0 (SDS HEARD ALL WAY DOWN TO ZERO MM HG)

S S S

162 MM HG

S S S S

S S S

S S S

0 MM HG

aortic insufficiency Continuous blood
) show wide pulse pressure with aortic
at sounds (S) are present from upper
o 0 mm



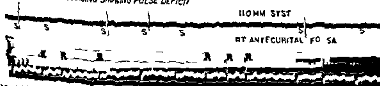


FIG. 657 Woman age 32 with rheumatic mitral stenosis and insufficiency and atrial fibrillati n. Upper tracing at apex hows accentuated first sound (S_1) systolic (SM) and diastolic (DM) murmurs Lower tracing a blood pressure tracing from the right antecu tal fossa. Note marked variation in intensity of sounds (S) some he e ard others particularly the rapid beats are not heard

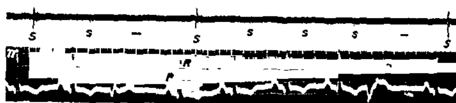


Fig. 658. A 63 year old man with coronary artery disease and frequent premature ventricular beats. Blood pressure *tracing*. Note absence of sound (—) made by premature beat and very loud sound after the compensatory pause.

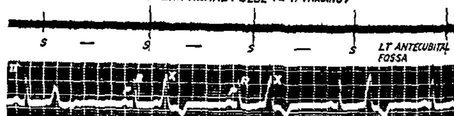


Fig. 659. Woman, age 48, with premature ventricular contractions (X) producing bigeminy. Blood pressure tracing, left antecubital fossa. Note absence (—) of sound (S) with each extrasystole (X).

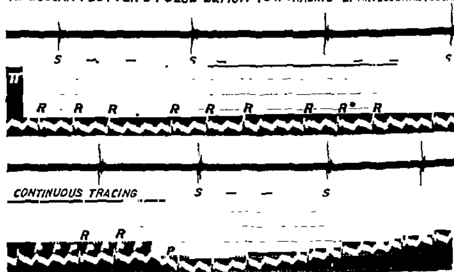


Fig. 660. A 67 year old man with paroxysmal atrial flutter. Continuous blood pressure tracing, left antecubital fossa Note absence of sounds (S) with more rapid ventricular contractions.

the average systolic or diastolic level (Fig. 657). We have been accustomed to denote as the systolic reading in cases of atrial fibrillation that point at which a significant number of beats are first heard, and as the diastolic reading the point at which almost all sounds become inaudible.

In the presence of occasional extrasystoles, the beat following the compensatory pause is generally stronger than the regular beats (Fig. 658). It is therefore logical to make determinations, whenever

possible, based on the sounds produced by the regular beats When there is constant bigeminy this is impossible The reading is then obtained from the stronger of the two beats and will necessarily be greater than if the rhythm were regular (Fig 659) In general all irregularities in rhythm such as atrial flutter (Fig 660) or varying heart block, may cause some confusion in blood pressure e timations We are aware that sound tracings obtained in this manner below the blood pressure cuff do no necessarily correspond with the palpability of the radial pulse Beats may not be audible that still can be felt at the wrist However this method was used because it adequately illustrated the points involved

Index

KEY TO ABBREVIATIONS USED IN THIS INDEX

S_1	First heart sound
S_2	Second heart sound
S_3	Third heart sound
DM	Diastolic murmur
PM	Presystolic murmur
SM	Systolic murmur

Page numbers in *italics* refer to illustrations